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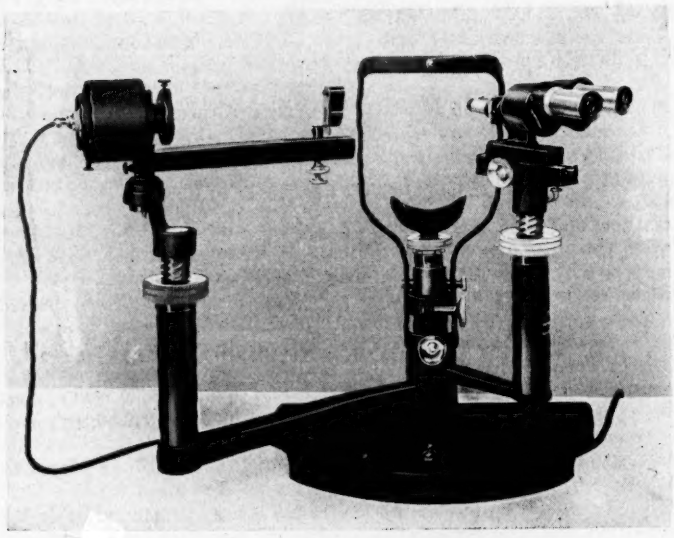

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Brit. J. Ophthal. (1958) **42**, 257.

COMMUNICATIONS

AMBLYOPIA ex ANOPSIA*

(SUPPRESSION AMBLYOPIA)

A PRELIMINARY REPORT OF THE MORE RECENT METHODS OF TREATMENT OF AMBLYOPIA, ESPECIALLY WHEN ASSOCIATED WITH ECCENTRIC FIXATION IN CASES OF STRABISMUS

BY

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DURING the last two centuries the treatment of amblyopia ex anopsia in cases of strabismus has made considerable progress. One of the earliest forms of treatment was occlusion of the fixing eye. This was first carried out by Buffon (1743), who also showed the need for correction of the refractive error by means of spectacles. More than a century later, Javal (1896) thought that squint was primarily a fault of binocular function to be remedied by orthoptic exercises using a stereoscope. Amongst others, Chavasse (1939) intensified the use of occlusion in the treatment of amblyopia and stressed the importance of early corrective operation in squint, and Lyle has continued the use of these methods (Lyle and Walker, 1953). It was found, however, particularly by Chavasse, that in certain cases, occlusion did not result in any appreciable improvement in vision, and that in others the improvement was minimal. Although it was known that some of these unresponsive patients showed eccentric fixation, treatment other than occlusion was not carried out until 1940, when Bangerter began to use what he described as "*pleoptic treatment*" (Bangerter, 1955), in an attempt to improve uniocular vision by means of exercises involving the coordination of hand, eye, and brain. This form of therapy is now becoming recognized as a successful means of treating patients who do not respond to occlusion. In 1953, Bangerter of St. Gallen, Switzerland, developed a special method of restoring foveal fixation. In 1954, Cüppers of Giessen, Germany, developed a different method. He constructed the Visuscope for the diagnosis of eccentric fixation and the Euthyscope for the restoration of central fixation (Cüppers, 1956). His after-image method of treatment of amblyopia has recently been introduced into England and a series of patients has been treated by this method at the High Holborn Branch of Moorfields Eye Hospital.

* Received for publication October 19, 1957.

Clinical Material

The patients chosen for treatment were between the ages of 4 and 13 years and were selected from among those with amblyopia ex anopsia who had not responded to occlusion of the fixing eye. In some of these cases occlusion had been carried out for between 1 and 8 months with no improvement in visual acuity. Many of them had been treated surgically and all had been classified as being satisfactory cosmetically. In some cases the diagnosis of eccentric fixation, or loss of central fixation, had already been made. The age at onset of the squint varied from birth to 3 years.

Instruments*

Visuscope.—This is used for diagnosis, and is essentially a conventional ophthalmoscope with low illumination. A small star is inserted in the path of light, the image of which is seen projected on to the patient's fundus.

Euthyscope.—This is used for treatment; it is also a modified ophthalmoscope but with high illumination. A beam of light is projected to form a cone of 30° of arc on to the posterior pole of the retina. In the centre of this cone there is an unilluminated area of either 3° or 5° of arc, as shown in the Figure (opposite). There is also a green filter which enables the examiner to locate the macula exactly before turning on the white light.

Coordinator.—This is used for treatment, and its action is based on the entoptic appearances of Haidinger's "brushes"—an effect produced by polarized light only visible to the patient when he is using the fovea, because of the special anatomical construction of that part of the retina.

Methods

Diagnosis.—The unaffected eye is kept firmly closed. The examiner, using the Visuscope, focuses the small black star onto the fundus of the affected eye, and the patient is instructed to look carefully at the star. The examiner then notes the exact position of the star on the fundus in relationship to the fovea centralis. The following types of fixation have been encountered:

(1) Central
  (*a*) With nystagmus;
  (*b*) Without nystagmus;
(2) Eccentric
  (*a*) Parafoveal and paramacular;
  (*b*) Peripheral; this may be found between the fovea and the disc, or even on the nasal side of the disc.
(3) Indefinite.

Treatment.—The first stage consists of total occlusion of the affected eye for a period of 3 to 4 weeks. When the fixation has become central, or is seen to be moving towards the fovea, the occlusion is changed to the unaffected eye, and pleoptic exercises are given. The after-image is produced with the Euthyscope, the pupil of the amblyopic eye being fully dilated. First the macula is found with the aid of the green filter, then the colour of the light is changed so that the macula is protected by the unilluminated area and a circular area of retina around the macula is stimulated; the light of the

* These instruments were all designed by Dr. C. Cüppers of Giessen, Germany.

Euthyscope, a 12- to 15-watt bulb, is projected on to the retina for 15 to 30 seconds; this results in the formation of an after-image corresponding to the area of retina stimulated. The after-image is either positive or negative depending on the intensity of suppression; it is usually positive at first and becomes negative later. The formation and observation of the after-image is facilitated by means of a flashing illumination on a white wall in the testing-room, the dark and light periods of the flashing being regulated independently.

The Coordinator is used for cases in which the fixation is near the macula and in which the after-image unavoidably covers the fovea as well as the eccentrically fixing point. It is also used in the later stages of the treatment of eccentric fixation after treatment has been begun with the Euthyscope. The patient is instructed to observe Haidinger's brushes and to bring and maintain them onto a fixation object controlled by his own hand. The Coordinator helps by showing the main projection of the macula outside the eye, and is therefore used to assist the restoration of foveal fixation, as well as to re-educate the coordination of eye, hand, and brain.

FIGURE.—Diagram of Euthyscope projecting a beam of light onto the fundus. (Courtesy of Clement Clarke Ltd.)

In-patients were treated daily and out-patients three times a week, each period of treatment lasting 30 minutes. Longer periods of treatment were found to be unsuitable because the patients became fatigued and no longer sufficiently cooperative.

Assessment of Results.—The patients under treatment for amblyopia were divided into two groups: those with central fixation, and those with eccentric fixation. After treatment had been completed, the improvement was assessed according to the visual acuity attained for distance :

(*a*) None, less than 6/36
(*b*) Slight, between 6/36 and 6/18
(*c*) Satisfactory, 6/12 or better.

The results were judged on the "cortical" rather than the "angular" visual acuity*. Near vision was judged separately and the improvement assessed as follows:

(*a*) None, less than N.36
(*b*) Slight, between N.36 and N.12
(*c*) Satisfactory, N.10 or better.

* The cortical visual acuity represents the ability to read a whole line of Snellen's test types accurately, whereas the angular visual acuity is the ability to read one letter per line. In suppression amblyopia, the angular visual acuity is usually better than the cortical.

The amount and duration of treatment depended on the degree of improvement obtained, and treatment was continued until it was clear that no further progress was likely to be made, or that further progress was likely to be so slow as to be uneconomic.

Results

(1) *Amblyopia with Central Fixation.*—There were twelve patients with amblyopia and central fixation (Table I, overleaf), eleven with convergent and one with divergent strabismus. All were between the ages of 5 and 13 years. When treatment was started the visual acuity for distance was between 6/18 and 6/36 (angular), and after treatment the results were as follows:

Improvement in Visual Acuity		Number of Cases
Distance	None	0
	Slight	2 (Visual acuity originally 6/36)
	Satisfactory	10
Near	None	0
	Slight	0
	Satisfactory	12

(2) *Amblyopia with Eccentric Fixation.*—There were 38 patients with amblyopia and eccentric fixation (Table II, overleaf), one with divergent, two with consecutive divergent*, and 35 with convergent strabismus. In the last group, two patients had a manifest vertical deviation, two a high degree of anisometropia, and thirteen a high degree of hypermetropia.

Of these 38 patients, 26 developed central fixation, and eight central fixation with nystagmus, and four remained with eccentric fixation. When treatment was started the visual acuity for distance varied between "the perception of hand movements" and 6/18 (angular), and after treatment the results were as follows:

Improvement in Visual Acuity		Number of Cases	
		Cortical	Angular
Distance	None	4	3
	Slight	16	8
	Satisfactory	18	27
Near	None	3	
	Slight	6	
	Satisfactory	29	

Before treatment by after-image stimulation was begun, occlusion of the unaffected eye had been carried out for an average period of about 8 months

* Consecutive divergent strabismus is a divergent squint resulting from an originally convergent squint, either due to over-liberal surgical correction, or to the mere passage of time.

for all but one of the cases with central fixation and all but seven of the cases with eccentric fixation, and no measurable improvement of visual acuity had resulted.

Discussion

The number of treatments necessary ranged from five to 53, the average being between twenty and thirty. On the whole less treatment was required for younger patients, particularly between the ages of 5 and 7 years, because they responded more quickly provided that their cooperation could be maintained. This is at variance with the opinion of Cüppers, who maintains that older children are more responsive to treatment. Patients with central fixation responded more quickly than those with eccentric fixation, and there was a more rapid response in those who were given daily treatment. The cases assessed as showing "no improvement" were mainly those in which there was a lack of cooperation on the part of the patient, or parent, in carrying out the treatment.

In those with central fixation, the after-image method of treatment rapidly overcame the suppression, although prolonged occlusion of the unaffected eye had previously failed to do so. In patients with eccentric fixation, the response to treatment appeared to depend upon the rapidity with which central fixation was regained; this in turn seemed to depend on the intensity of the macular suppression. As soon as the fixation became central, the angular visual acuity rapidly improved and total occlusion of the unaffected eye was carried out. This increased the rate of improvement of visual acuity, which showed a greater improvement in all cases if measured when the after-image was still present. This is said to indicate the degree to which the angular vision may be expected to improve, given sufficient time (Cüppers).

The cortical vision, especially for distance, was invariably slower to improve than the angular vision, and it was also observed that near vision improved to a greater extent than distance vision. This was thought to be due to the extensive use of Haidinger's brushes and the fact that the homework carried out was concerned with near vision, *e.g.* exercises such as crossing out all the letters "e" in a page of newsprint, as well as reading, writing, and drawing. It has been suggested that distance vision shows less improvement because children are not particularly handicapped with a visual acuity of 6/12 or 6/18, and therefore make less effort to see clearly in the distance.

Until recently it was believed that, in some cases, the visual acuity could not be improved beyond the point to which it had developed at the time of onset of the inhibition caused by the commencement of the squint. The amblyopia, represented by the visual acuity, which had developed at the time of onset of the squint, is known as the amblyopia of arrest (Chavasse).

This series included two cases in which the squint was known to have begun in the first few months of life (Cases 8 and 10 in Table II). It is

TABLE
RESULTS IN TWELVE CASES OF AMBLYOPIA EX AN
TREATED BY CÜPPERS'S AFTER

Case No.	Age (yrs)	Diagnosis	Age at Onset of Strabismus (yrs)	Previous Treatment	No. of Treatments	Fixation of Affected Eye		Before
						Before Treatment	After Treatment	Distance
1	6	Right convergent strabismus Abnormal retinal correspondence	4/12	Yes	10	Central with nystagmus	Central	6/36
2	8	Right convergent strabismus High hypermetropia*	From birth	Yes	11	Central	Central	6/18
3	5	Left convergent strabismus Abnormal retinal correspondence Left hypertropia Myopia	15/12	Yes	8	Central with nystagmus	Central	6/36
4	9	Intermittent right divergent strabismus Myopia	5	Yes	12	Central with nystagmus	Central	6/24
5	6	Left convergent strabismus Abnormal retinal correspondence	4	Yes	19	Central with nystagmus	Central	6/24
6	6	Left convergent strabismus Abnormal retinal correspondence	9/12	Yes	5	Central with nystagmus	Central	6/36
7	9	Left convergent strabismus Abnormal retinal correspondence High hypermetropia	2	Yes	5	Central	Central	6/36
8	5	Right convergent strabismus Abnormal retinal correspondence	3	Yes	10	Central	Central	6/24
9	8	High hypermetropia	6	Yes	28	Central	Central	6/36
10	7	Left convergent strabismus	4	Yes	10	Central with nystagmus	Central	6/24
11	13	Esophoria Intermittent convergent strabismus Myopia	From birth	No	6	Central with nystagmus	Central	6/24
12	13	Left divergent strabismus Myopia	6	Yes	13	Central with nystagmus	Central	6/36

* High hypermetropia = more than +3D sph.

suggested that the visual acuity did not improve as a result of occlusion of the unaffected eye because there was an undiagnosed eccentric fixation, which the occlusion had only served to encourage. The vision, therefore, had only improved to the extent that the resolving power of the eccentrically fixing point would allow, which is usually not better than 6/24. In these cases diagnosis by the Visuscope revealed an eccentrically fixing point on the retina. After the restoration of foveal fixation, by the after-image treatment, the visual acuity subsequently improved to 6/6 angular, and binocular single vision, with some range of fusion, was obtained. Cheiroscope drawing

TABLE
LYOPIA
PPERS'S

EX ANOPSIA WITH CENTRAL FIXATION
AFTER-IMAGE METHOD

Affected	After Treatment	Visual Acuity of Affected Eye						Binocular Function
		Before Treatment		After Treatment				
		Distance	Near	With After-Image	Angular	Cortical		
						Distance	Near	
	Central	6/36	N.36	6/6	6/9	6/12	N.5	Can alternate
	Central	6/18	N.18	6/4·5	6/7·5	6/9	N.5	Fusion and stereoscopic vision Straight on cover test
	Central	6/36		6/4·5	6/6	6/6	N.5	None
	Central	6/24	N.24	6/6	6/9	6/12	N.5	Binocular single vision for near and distance
	Central	6/24	N.14	6/4·5	6/6	6/12	N.5	None
	Central	6/36		6/6	6/12	6/18	N.6	Cannot alternate
	Central	6/36	N.24	6/4·5	6/6	6/6	N.5	Can alternate
	Central	6/24		6/4·5	6/6	6/12	N.5	Cannot alternate
	Central	6/36	N.10	6/12	6/12	6/12	N.5	Fusion and stereoscopic vision Straight on cover test
	Central	6/24		6/6	6/9	6/12	N.5	None
	Central	6/24	N.24	6/6	6/6	6/6	N.5	Binocular single vision
	Central	6/36	N.36	6/12	6/12	6/18	N.8	Binocular single vision for near

confirmed that this was true fusion.

Better and quicker results were obtained in those patients who had not had any previous occlusion, even when the fixation was sufficiently eccentric to be diagnosed by the cover test alone. Therefore, it is recommended that, in any case of amblyopia, a diagnosis of the type of fixation present should be made before occlusion, or any other form of treatment, is carried out.

The results obtained in this series are encouraging and it is hoped that the treatment will be continued. When this type of treatment is contemplated, it is desirable that it should be given as soon as the patient has developed the

TABLE II

RESULTS IN 38 CASES OF AMBLYOPIA EX ANOPSIA TREATED BY CÜPPERS' AFTER-

Case No.	Age (yrs)	Diagnosis	Age at Onset of Strabismus (yrs)	Previous Treatment	No. of Treatments	Fixation of Affected Eye		
						Before Treatment	After Treatment	Before Distance
1	7	Left convergent strabismus Lack of normal retinal correspondence	8/12	Yes	21	Between disc and fovea	Central with nystagmus	5/60
2	10	Left convergent strabismus Abnormal retinal correspondence High hypermetropia*	6	Yes	36	Nasal border of macula	Central	6/36
3	7	Left convergent strabismus Abnormal retinal correspondence High hypermetropia	1½	Yes	36	Nasal border of macula	Central	6/24
4	6	Right convergent strabismus Abnormal retinal correspondence High hypermetropia	3	No	21	Between disc and macula	Central	Counting fingers a 30 cm.
5	7	Left convergent strabismus Normal retinal correspondence	4	Yes	8	Nasal border of macula	Central	6/24
6	8	Left convergent strabismus Abnormal retinal correspondence	1	Yes	14	Nasal border of macula	Central	6/36
7	6	Right convergent strabismus Anisometropia	1	No	12	Nasal to the fovea	Central	5/60
8	5	Right convergent strabismus Abnormal retinal correspondence	5/12	No	10	Between disc and fovea	Central	1/60
9	4	Left convergent strabismus Abnormal retinal correspondence	Uncertain	No	12	Between disc and fovea	Central with nystagmus	1/60
10	5	Left convergent strabismus Abnormal retinal correspondence High hypermetropia	3/12	Yes	16	Nasal to fovea	Central	6/60
11	5	Left convergent strabismus Abnormal retinal correspondence High hypermetropia	1	Yes	8	Nasal border of fovea	Central	6/36
12	7	Left convergent strabismus Abnormal retinal correspondence Left hypertropia	From birth	Yes	20	Nasal to fovea	Central with nystagmus	6/36
13	10	Right convergent strabismus Abnormal retinal correspondence High hypermetropia	From birth	Yes	35	Nasal border of disc	Parafoveal	Counting fingers at 1 m.
14	5	Right divergent strabismus	8/12	Yes	16	Temporal to fovea	Central	6/36
15	6	Left convergent strabismus Lack of normal retinal correspondence High hypermetropia	From birth	No	28	Temporal to disc	Central nystagmus	Hand movements

* High hypermetropia = more than +3D sph.

TABLE II
AMBLYOPIA EX ANOPSIA WITH ECCENTRIC FIXATION
(UPPERS' AFTER-IMAGE METHOD

Visual Acuity of Affected Eye						Binocular Function
Before Treatment		After Treatment				
				Cortical		
Affected	After Treatment	Distance	Near	With After-Image	Angular	Distance	Near	
Central w nystagm		5/60	None	6/18	6/24	6/60	N.24	None
Central		6/36	N.36	6/4·5	6/9	6/12	N.5	Can alternate
Central		6/24	N.10	6/6	6/9	6/12	N.5	Fusion and stereoscopic vision
Central		Counting fingers at 30 cm.	None	6/18	6/36	6/36	N.6	None
Central		6/24	N.24	6/9	6/9	6/12	N.5	Weak fusion No stereoscopic vision Seems straight on cover test
Central		6/36	N.24	6/6	6/9	6/12	N.5	Can alternate
Central		5/60	None	6/12	6/12	6/18	N.5	Can alternate for near
Central		1/60	None	6/6	6/6	6/6	N.5	Fusion and stereoscopic vision
Central w nystagm		1/60	None	6/24	6/36	6/60	N.18	None Attendance insufficient
Central		6/60	None	6/5	6/6	6/6	N.5	Fusion and stereoscopic vision Straight on cover test
Central		6/36		6/6	6/12	6/18	N.5	Can alternate
Central w nystagm		6/36	N.10	6/4·5	6/18	6/18	N.5	Abnormal retinal correspondence
Parafove		Counting fingers at 1 m.	None	6/36	4/60	4/60	None	None
Central		6/36	None	6/6	6/9	6/9	N.5	Simultaneous macular perception
Central nystagm		Hand move-ments	None	6/18	6/36	6/60	N.36	None Lack of normal retinal correspondence

Continued—

TABLE II (continued)

Case No.	Age (yrs)	Diagnosis	Age at Onset of Strabismus (yrs)	Previous Treatment	No. of Treatments	Fixation of Affected Eye		Before T
						Before Treatment	After Treatment	Distance
16	10	Left convergent strabismus Abnormal retinal correspondence High hypermetropia	?4	Yes	41	Nasal to fovea	Central	6/24
17	10	Left convergent strabismus Abnormal retinal correspondence High hypermetropia	2	Yes	32	Parafoveal with nystagmus	Central	6/24
18	7	Left convergent strabismus Abnormal retinal correspondence	Uncertain	No	39	Nasal to fovea	Central	5/60
19	12	Right convergent strabismus	4	Yes	28	Nasal to fovea	Central	6/18
20	9	Right convergent strabismus Abnormal retinal correspondence High hypermetropia	1½	Yes	32	Temporal to fovea	Central with nystagmus	6/36
21	5	Right convergent strabismus Abnormal retinal correspondence High hypermetropia	1	No	27	Between disc and fovea	No real fixation	Counting fingers at 50 cm.
22	6	Right convergent strabismus Abnormal retinal correspondence Right hypertropia	16/12	Yes	25	Nasal to fovea	Central	6/36
23	5	Right convergent strabismus Abnormal retinal correspondence	?3	Yes	7	Nasal to fovea Occasionally central	Central	6/36
24	7	Left convergent strabismus Abnormal retinal correspondence	?	Yes	33	Nasal border of fovea	Central	6/60
25	8	Left convergent strabismus Abnormal retinal correspondence	2	Yes	38	Nasal border of fovea	Central	6/24
26	8	Left convergent strabismus Abnormal retinal correspondence	1	Yes	34	Nasal border of fovea	Central	6/24
27	6	Right convergent strabismus Abnormal retinal correspondence	13	Yes	8	Nasal border of fovea	Central with nystagmus	6/36
28	9	Left convergent strabismus Abnormal retinal correspondence	?	Yes	15	Nasal border of fovea	Central	6/36
29	7	Left convergent strabismus Abnormal retinal correspondence	?	Yes	12	Nasal border of fovea	Central	6/24
30	10	Right convergent strabismus Abnormal retinal correspondence Anisometropia +5D astigmatism	2	Yes	53	Between disc and fovea	Central	1/60

TABLE III (*continued*)

Visual Acuity of Affected Eye						Binocular Function
Before Treatment		After Treatment				
				Cortical		
Distance	Near	With After-Image	Angular	Distance	Near	
6/24	N.36	6/4·5	6/9	6/12	N.5	Abnormal retinal correspondence
6/24	N.24	6/4·5	6/9	6/12	N.5	Superimposes slides First grade binocular vision only
5/60	None	6/6	6/12	6/24	N.8	Cannot alternate
6/18	N.18	6/4·5	6/6	6/9	N.5	Fusion poor
6/36	N.24	6/6	6/12	6/24	N.6	None
Counting fingers at 50 cm.	None	6/30	6/60	6/60	N.36	None
6/36	N.24	6/9	6/9	6/12	N.5	None
6/36	N.36	6/9	6/9	6/18	N.5	Can alternate
6/60		6/4·5	6/6	6/12	N.5	Diplopia caused by treatment for abnormal retinal correspondence
6/24	N.24	6/6	6/9	6/12	N.6	Binocular single vision for near
6/24	N.18	6/4·5	6/12	6/18	N.5	Can alternate
6/36		6/12	6/12	6/18	N.6	Can alternate
6/36	N.18	6/6	6/6	6/6	N.5	None
6/24		6/4·5	6/9	6/12	N.5	Fusion
1/60	None	6/9	6/24	6/36	N.24	Slight fusion

Continued—

TABLE II (continued)

Case No.	Age (yrs)	Diagnosis	Age at Onset of Strabismus (yrs)	Previous Treatment	No. of Treatments	Fixation of Affected Eye		Before T
						Treatment Before	After Treatment	Distance
31	12	Left divergent strabismus (consecutive) High hypermetropia	5/12	Yes	47	Temporal to fovea	Central wi nystagm	5/60
32	6	Left convergent strabismus Abnormal retinal correspondence	1	Yes	5	Nasal border of fovea	Central	6/24
33	9	Left convergent strabismus Abnormal retinal correspondence High hypermetropia	2	Yes	31	Nasal to fovea	Central	6/36
34	13	Right divergent strabismus (consecutive)	2	Yes	37	Temporal to macula	Central occasion ally	2/60
35	10	Left convergent strabismus Lack of normal retinal correspondence	?4	Yes	22	Nasal border of fovea	Central	6/36
36	6	Left convergent strabismus Abnormal retinal correspondence	6/12	Yes	6	Nasal border of fovea	Central	6/24
37	7	Right convergent strabismus Abnormal retinal correspondence	From birth	Yes	38	Nasal to disc	Central (unsteady	3/60
38	8	Left convergent strabismus Abnormal retinal correspondence	1	Yes	18	No real fixation in fovea	Central wi nystagm	6/24

power to cooperate, in order to provide him with a high standard of visual acuity in each eye at the earliest possible age. Thus, he will have a reserve eye if he ever has the misfortune to lose the vision of one eye through trauma or some organic lesion. The probability of subsequent deterioration of vision in the originally amblyopic eye must, however, be borne in mind. Only in those cases in which restoration of binocular single vision, or some form of subnormal binocular vision, has occurred, or in those rendered freely alternating, can it be certain that there may not be some subsequent deterioration of the visual acuity gained in this way. However, if adequate treatment is given, it is unlikely that there will be a return to eccentric fixation once central fixation has been established, and it is unlikely that the visual acuity developed will later become grossly reduced. Follow-up examination of these cases will be needed in order to find this out.

Summary

Fifty cases of amblyopia were treated by Cüppers's after-image method; 38

TABLE II (*continued*)

Visual Acuity of Affected Eye						Binocular Function
Before Treatment		After Treatment				
				Cortical		
Distance	Near	With After-Image	Angular	Distance	Near	
5/60		6/12	6/18	6/24	N.14	Fusion Binocular single vision at times
6/24	N.10	6/6	6/9	6/9	N.5	None
6/36	N.24	6/6	6/12	6/18	N.6	Can alternate
2/60		6/24	6/60	6/60	N.24	None
6/36	N.24	6/4·5	6/9	6/12	N.5	First grade binocular vision only
6/24	N.24	6/4·5	6/6	6/6	N.5	Can alternate
3/60	None	6/9	6/36	6/36	N.36	None
6/24		6/9	6/9	6/12	N.5	Can alternate

showed eccentric fixation in the amblyopic eye and the remaining twelve had central fixation. The vision in 38 cases improved to 6/12 and N.10 or better after treatment.

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MODIFIED MAJOR AMBLYOSCOPE*

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IN the assessment of a patient with comitant strabismus, it is essential to determine the angle between the fixation axes ("objective" angle of squint), the state of the retinal correspondences, the angle of binocular projection ("subjective" angle of squint), and the grade of binocular vision. All methods of determining these factors have their limitations.

Angle between Fixation Axes ("Objective" Angle of Squint)

The most commonly used methods of determining this angle depend on abolishing any ocular movement when fixation changes from one eye to the other. In using the major amblyoscope for this purpose, a fixation object is supplied alternately to each eye, the arms of the instrument being adjusted until no ocular movement takes place. There is thus complete dissociation between the eyes, and the conditions are far from being those obtaining in the normal use of the eyes; moreover, even though the images are optically at infinity, they often appear to be suspended in space at a short distance from the observer. In using Duane's parallax test for measuring the angle, alternate occlusion is again employed, with a prism in front of one or both eyes, so that again the conditions are unusual. It is not surprising that the angle measured by these methods often differs from that suggested by the patient's appearance, by the position of the corneal reflexes, and by the behaviour on cover test.

Methods depending on the corneal reflex may be carried out under more normal conditions of viewing, but are in general either inaccurate or clumsy, and require an independent measurement of the angle gamma. A method which removes some of these disadvantages has been described by Graham and Naylor (1957).

State of the Retinal Correspondences

The three most useful methods of investigating the state of the correspondences (Duke-Elder, 1949) are the after-image test, the congruence test of Tschermak, and the major amblyoscope. The conditions of viewing differ

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greatly from test to test, and in none do they bear a direct relationship to everyday life, so that the state of retinal correspondence elicited may vary from test to test in the same patient, and also from time to time.

Angle of Binocular Projection (*"Subjective" Angle of Squint*)

If the retinal correspondences are normal, this angle is the same as the angle between the fixation axes; if the retinal correspondences are abnormal, the difference between the two angles is the angle of anomaly. The angle of binocular projection can be measured in several ways, all of which depend on dissociating the images of the two eyes; this may be done by excluding the field of one eye from the other (mechanically as in cover tests, Maddox Wing test, major amblyoscope, etc.; by colour as in the red-green tests; or by polarized light), by displacing one image and so inducing diplopia (*e.g.* prism diplopia test and the various phorometers), or by utilizing after-images. The results again vary from test to test and from time to time, and again, since the fields of vision are dissociated, the conditions are far from being those of everyday life.

Grade of Binocular Vision

This is usually assessed by means of a major amblyoscope, which allows accurate assessment of the binocular vision under its own conditions of viewing, but does not in itself indicate how much use is made of binocular vision in everyday life. Interrupted reading, such as by bar reading or the Bishop Harman diaphragm, gives a very useful indication of binocular function, but even in these tests the vital stimulus is a monocular one; and the colour tests, such as the Friend test and Worth's four-dot test, indicate the use of binocular vision only under the special conditions of each test.

In general, the cover test gives by far the most useful information; although it does not give a direct estimate of the grade of binocular vision, it does indicate directly the position of the eyes under physiological conditions. When it gives an unequivocal result, it is usually sufficient, in combination with major amblyoscope readings, for an adequate assessment to be made. In some circumstances, however, assessment may not be so easy:

(*a*) If the patient's attention wanders, because of youth or slow mental development, fixation may not be maintained sufficiently long for an adequate assessment to be made. This is particularly so in cases with a small angle of squint and with fixation disparity.

(*b*) In some patients with heterophoria, the cover test itself may produce a breakdown of compensation.

(*c*) Since the cover test does not give a direct estimate of the grade of binocular vision present, it may not indicate exactly why the eyes take up a certain position. For example, if the eyes appear to be straight for one distance only, particularly after surgical treatment and in the presence of equivocal major amblyoscope

readings, it may be uncertain whether the eyes are straight because of central fusion or of peripheral fusion only, or because of particularly accurate surgery without the help of the patient's binocular vision. The interpretation of small degrees of esotropia and fixation disparity can also be very difficult (Gittoes-Davies, 1952; Jampolsky, 1956).

There appears, therefore, to be a place for an instrument on which the various measurements can be carried out under conditions approaching those of normal life.

Modified Major Amblyoscope

The general construction of the instrument (Fig. 1) is that of the routine major amblyoscope, with all the usual adjustments and provision for an after-image test. The end of each arm nearer to the patient, carrying the reflector and lens and eyepiece, is made so that it is detachable from the main body of the tube (Fig. 2, opposite) by means of a slide having bevelled vertical edges.

Two alternative fittings are provided for each arm: one carries the usual opaque reflector, lens, and eyepiece to provide for routine measurements in the usual way, and the other provides for the modified method of measurement. The transfer from one fitting to the other, consisting only of sliding in one end-piece on each arm in place of another, takes a few seconds only. The fittings not in use may be stored in dovetailed slides on the stand of the instrument (as in Fig. 1) or in a separate box.

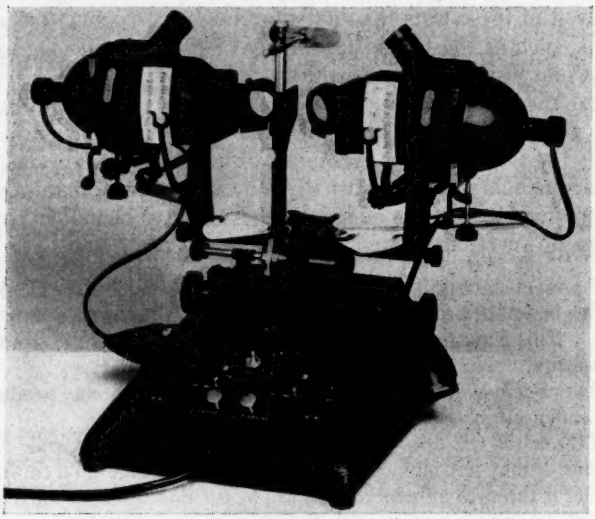

Fig. 1.—General view of modified major amblyoscope.

FIG. 2.—Interchangeable fittings for routine and modified methods of examination.

The principle of the modified instrument is shown in Fig. 3. The opaque mirrors incorporated in each arm of the routine major amblyoscope are replaced by transparent mirrors M, so that the patient can see through them with both eyes simultaneously to a target T at a distance, thus providing a natural binocular stimulus. At the same time pictures can be projected on to each retina from the slide-holders H in each arm, the face of the transparent mirror M nearer to the patient acting as the main reflecting surface. A fainter reflection is also produced by the back surface of the mirror, but trouble from this is avoided by inclining this surface relative to the front one, so that the two images virtually coincide; the prismatic effect thus introduced is far too small to interfere in any way with the measurements. The lens

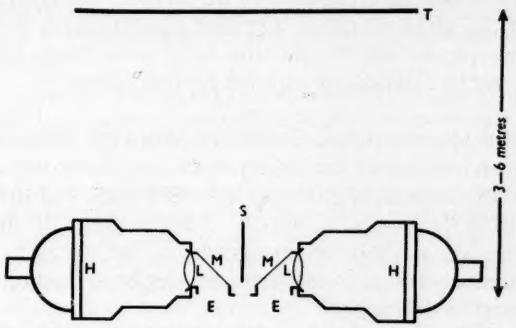

FIG. 3.—Principle of modified major amblyoscope (for lettering see text).

which is normally incorporated in the eyepiece E, and which serves to put the image of the slide optically at infinity, is replaced by a +8D lens L placed between the slide and the mirror, serving the same purpose but not obstructing the patient's view of the target. A septum S prevents troublesome reflections of the light from the opposite arm of the instrument. The patient's subjective impression is of an uninterrupted wide-angle (35°) view of a distant target, superimposed on which he sees the images of the slides. The conditions of viewing approximate closely to those of everyday life.

The target T used in most measurements is a patterned wall or screen (wallpaper is very convenient) executed preferably in greys or fawns so that it does not make the images of the amblyoscope slides too difficult to distinguish when they are superimposed upon it, but with a sufficiently bold and well-defined design to give adequate stimulus to fusion. It is best placed at 6 metres from the instrument for routine use, but may be nearer when it is desired to assess visual functions for shorter distances. If the space for the full 6 metres is not available, perfectly satisfactory results can be obtained at shorter distances, and most of the patients described in this paper have been measured at 3 metres; at these shorter distances it must be remembered that the eyes will be slightly convergent when they are being used normally, and this must be allowed for; at 3 metres, the normal scale reading on the amblyoscope will be +1°. This difference will not be of great importance in most patients, unless measurements of fixation disparity are being made (see below). When the eye is focused for these shorter distances, the image of the slides, which are optically at infinity, will not be perfectly in focus, and theoretically the power of the lens L should be changed; if the working distance is fixed, this can easily be done during the construction of the instrument, but in any case the slight fuzziness at 3 metres ($\frac{1}{3}$ dioptre error) produced by an instrument designed for 6 metres does not interfere in any way with its use.

The slides are identical in nature with those used routinely with the major amblyoscope, except in two respects; in the first place, the pictures are executed on a black background, so that, when projected in the instrument, only the picture itself is seen superimposed on the target. Secondly, the change in the position of the lens L results in a greater magnification than in the usual major amblyoscope, so that the pictures have to be made correspondingly smaller if they are to correspond with the routine slides.

MEASUREMENT OF OBJECTIVE AND SUBJECTIVE ANGLES.—Since the slides used with the modified instrument are largely black and allow only a little light to reach the eye, the corneal reflections cannot be seen and are therefore no guide to the objective angle of squint. Measurement of this angle must therefore depend on abolition of movement of the eye as fixation changes from one eye to the other, and can therefore only be carried out if monocular fixation is central in both eyes.

Using slides similar to the routine S.P. or S.M.P. slides, the flashing buttons

are operated so that only one eye (the fixing eye) is presented with a picture from the slide; the arm of the instrument is adjusted so that the image is seen roughly in the centre of the distant target which is, of course, seen by both eyes. The second arm of the instrument is then adjusted until a change of fixation to the second eye (produced by operating the flashing buttons so that only the second eye sees the image of a slide) produces no movement of this eye. During the measurements it is essential to make sure that the patient's eyes are close to the eyepiece; if the head is allowed to move from this position, the view of the target is restricted and does not provide an adequate stimulus. This may make it difficult to watch the eyes over the top of the eyepieces as with the routine instrument, but they can still be seen by looking through the upper part of the transparent mirror; this can easily be done without obstructing the patient's view of the distant target.

The subjective angle is also measured with S.P. or S.M.P. slides, one of which is presented as before to the fixing eye with the arm adjusted so that it is seen in the centre of the target. The patient is then asked to move the second arm of the instrument until the images of the slides are superimposed.

In both these measurements, the distant target is seen continuously by both eyes, which therefore take up the positions occupied in everyday life; the images of the slides act merely as markers which indicate the objective and subjective angles and do not influence the positions of the eyes. This is particularly obvious in patients with esophoria, who give readings indicating no deviation of the eyes on the modified instrument.

In all these measurements, it is essential to have the target well illuminated, and to adjust the illumination of each slide individually so that there is only just sufficient light to make the picture visible against the target background and the strongest visual stimulus is still the distant target; if the slide illumination is too high or the target illumination too low, the slide pictures become the predominant stimulus and tend to be seen in front of the target. In this way the whole principle of the method is lost, as in the following example:

Case 1, a boy aged 10 years, had had a right convergent comitant squint with equal visual acuity since the age of 6 years, with some fusion and weak ductions but a marked tendency to right eye suppression.

Examination.—The cover test showed a manifest right convergent squint with and without glasses most of the time, but occasionally he appeared to be straight for distance with glasses. A recession of the right medial rectus muscle had been performed at age of 8 years.

When he was seen for assessment, routine major amblyoscope readings showed S.P. and fusion at +20°, with ductions from +16° to +30°; the eyes looked quite straight, but binocular vision broke down rapidly on cover testing to show a convergent squint. The modified amblyoscope gave an angle of +20° with S.P. slides when the slide illumination was relatively high and the light in the room (and therefore the target illumination) rather dim, but he showed no deviation when the room light was increased and the slide illumination reduced to a minimum.

Similarly, the slides carrying small pictures (S.M.P. type) often give more reliable information than larger pictures (S.P. type), since the larger image of the latter may tend to be dominant over the peripheral fusion stimulus given by the background target, so that the readings approximate more closely to those of the routine method, as in Cases 2 and 3.

Case 2, a girl aged 5 years, had an intermittent divergent squint of the divergence excess type.

Examination.—Routine amblyoscope measurements showed S.M.P. and fusion at $-10°$, with ductions to $+5°$; the modified instrument showed S.M.P., fusion, and S.V. at 0°, but S.P. slides were superimposed at $-8°$.

Case 3, a girl aged 7 years, had had a left accommodative convergent squint with partial left amblyopia since the age of 3 years; after occlusion, the visual acuity was 6/9 in each eye. A cover test with glasses showed a variable degree of squint, sometimes definitely straight for distance and near; without glasses, she had a manifest left convergent squint for distance, but no deviation for near.

Examination.—Routine major amblyoscope readings showed fusion at $+5°$ with suppression on ductions. The modified instrument at 3 metres consistently gave readings indicating no deviation with S.M.P. slides, but $+8°$ to $+12°$ with S.P. slides.

S.M.P. slides or the smallest practicable slides should therefore always be used.

Measurements of the objective and subjective angles may give results comparable with those obtained with the routine major amblyoscope, but in a number of patients the angle on the modified instrument was appreciably different, as in the following example:

Case 4, a boy aged 7 years, had had an alternating convergent comitant squint, with a rather variable angle, since the age of 6 months. He had had a resection of the right lateral rectus and recession of the right medial rectus muscles 3 months previously.

Examination.—The post-operative objective angle on the routine major amblyoscope was $+20°$, but on the modified instrument only $+13°$. No subjective angle could be obtained on either instrument.

Since a different amount of surgery is indicated for angles of $+20°$ and $+13°$, this difference is obviously of significance.

A limitation of this modified method of examination is that measurements of angles of more than 25° cannot be taken accurately; this is because the arm of the instrument corresponding to the fixing eye must remain stationary in such a position that this eye sees the image of the slide in the centre of the target, and as the second arm is moved inwards it gradually cuts off the view of the target by the non-fixing eye. This is probably of relatively little importance; in the first place, the angle between the two eyes in such patients is probably determined mainly by the conditions presented to the fixing eye rather than to the non-fixing eye, and the field of vision of the fixing eye is not affected by the movement of the other arm of the instrument; in the second place, differences in the readings given by the modified and routine methods of measurement are of little practical importance with large angles of squint, since they are unlikely to influence the amount of surgery to be performed.

STATE OF THE RETINAL CORRESPONDENCES

If the subjective and objective angles are equal, the retinal correspondences are normal; if they are unequal, the difference is the angle of anomaly. It has already been pointed out that the apparent state of the retinal correspondences may depend on the type of test employed, and the presence or absence of abnormal retinal correspondences (A.R.C.) on routine testing on the major amblyoscope does not necessarily indicate that the same result would be obtained under normal viewing conditions. This is indicated by the following example; in this patient the modified instrument indicated abnormal retinal correspondence long before it was detected on the routine instrument:

Case 5, a boy aged 7 years, had had a left convergent squint with marked amblyopia since the age of 1 year; occlusion had restored equal visual acuity.

Examination.—On routine testing in the orthoptic department, he showed an angle of +35° with S.P. only, and A.R.C. had not been suspected at any time. The modified instrument at 6 metres showed an objective angle of about +30°, but a subjective angle of 0° with S.P. slides. Resection of the left lateral rectus and recession of the left medial rectus muscles were then performed; after this no binocular vision could be demonstrated on routine testing for 12 months, but 2 years later the routine tests on the major amblyoscope showed A.R.C., with an objective angle of +2° and subjective angle of −5°.

Occasionally patients who have no binocular vision on routine major amblyoscope measurements can be shown by the modified method to have abnormal retinal correspondence:

Case 6, a girl aged 12 years, had had a left convergent squint with paresis of the left superior oblique muscle and bilateral overaction of the inferior oblique muscles since the age of 2 years. Intermittent occlusion had been required for partial left amblyopia, the final visual acuity being 6/5 in the right and 6/12 in the left. When she was 11 years old, a bilateral myectomy of the inferior oblique muscle was performed, followed by lateral rectus resection and medial rectus recession in both eyes.

Examination.—When she was seen post-operatively for assessment, the cover test showed a small manifest left convergent squint for near and distance. Routine major amblyoscope readings showed an objective angle of +7°, and measurements on many occasions had failed to demonstrate S.P. On the modified instrument, the objective angle was +6°, and the subjective angle was −2° with large S.P. slides and 0° with S.M.P. slides, though the latter showed a marked tendency to left eye suppression.

Case 7, a boy aged 14 years, had had a left convergent squint with partial left amblyopia since the age of 5 years. After occlusion, the final visual acuity was 6/6 part in the right eye and 6/9 in the left. When he was 13 years old, a bilateral resection of the lateral recti had been performed.

Examination.—When he was seen for final assessment, glasses were not worn; the routine major amblyoscope showed no S.P., though on previous occasions attempts at S.P. had been made; the modified instrument showed an objective angle of +9° and a subjective angle of 0° with both S.P. and S.M.P. slides.

Similarly, a non-harmonious A.R.C. found on routine testing may be shown to be harmonious in normal viewing conditions:

Case 8, a girl aged 9 years, had had a manifest right convergent squint and paresis of the

right lateral rectus muscle since the age of 4 years, with partial amblyopia (R.V. 6/36. L.V. 6/9), and nystagmoid movements of the right eye on abduction. On routine testing, the objective angle fixing right was +20° L/R 9△, and fixing left +20° L/R 3△, and she attempted S.P. at +5° L/R 4△ when fixing with the right eye. With the modified instrument, the subjective angle was 0° with no height, the objective angle fixing right and fixing left being +10°/+15°. A resection of the right lateral rectus muscle was performed; 2 months later the objective angle on the modified instrument was 0°, the subjective angle being −5° with S.P. slides and −10° R/L 3△ with S.M.P. slides.

Harmonious A.R.C. may occasionally be demonstrated in patients who are incapable of doing subjective tests on the routine instrument:

Case 9, a girl aged 5 years, had had a right convergent comitant squint for 1 year for near and distance, with and without glasses, with partial amblyopia (visual acuity 6/18, in the right eye and 6/12 in the left, after occlusion).

Examination.—The angle of squint was very variable, but appeared to be between +10° and +30°; she was too young for subjective readings on the routine major amblyoscope, but gave consistently a subjective angle of 0° on the modified instrument.

It is noteworthy that, in all these patients, the abnormal correspondence under normal viewing conditions was harmonious at the original tests, though in Case 8 it became unharmonious immediately after operation. There appears to be a definite tendency for A.R.C. to be harmonious in normal viewing conditions more commonly than would be expected from the routine readings on the major amblyoscope; this was also found to be the case by Halldén (1952) and Levinge (1953). Nevertheless, some patients have unharmonious A.R.C. even on the modified instrument:

Case 10, a boy aged 7 years, had had a manifest alternating convergent comitant squint since the age of 2 years; the onset was associated with four or five mild epileptic fits.

Examination.—The objective angle before operation was +20° with glasses, +40° without glasses, with marked alternating suppression. A bilateral resection of the lateral recti had been performed 3 months before he was seen for assessment. On examination, the objective angle was +14° both on the routine major amblyoscope and on the modified instrument; no subjective reading was possible on the routine instrument, but on the modified instrument the subjective angle with S.P. slides was consistently +5°.

In this patient, the recent operation may again be responsible for the failure of the A.R.C. to be harmonious; if that were the case and the A.R.C. had been present before operation, however, it would be expected that the angle of anomaly after the operation would have been greater rather than less than the angle of squint.

Grade of Binocular Vision

The binocular vision being used under normal viewing conditions can be assessed in several ways:

(*a*) Unless the readings with the modified instrument indicate no objective or subjective deviation of the eyes, the patient cannot be exercising normal binocular vision. If there is no objective or subjective deviation, this may be the result of

central fusion, peripheral fusion, or possibly of accurate surgery without useful binocular vision.

(*b*) When using S.P. or S.M.P. slides, central suppression is manifest in the same way as on the routine instrument.

(*c*) *In patients with no deviation, or with only a small angle of squint, fusion is* best tested as follows:

The arms of the instrument are locked in the position indicating no ocular deviation (*i.e.* at 0° for a distant target, +1° for a target at 3 metres). S.P. or S.M.P. slides are used, depending on the amount of central suppression present; if no deviation is present, the patient will of course see them superimposed on the target, but if a squint is present and the retinal correspondences are normal, the pictures will appear separated. In the latter case, prisms are introduced in the eyepiece usually of the non-fixing eye, until the pictures are superimposed; prisms of both greater and less power than this are then introduced, using both eyepieces if necessary, and the range of powers over which the slides remain superimposed is the measure of the amplitude of fusion. The principle of this method is shown in Fig. 4. The image of the distant target, and the images of the slides from the arms set in the position corresponding to no deviation, are all affected equally by the prisms P in the eyepieces; provided the eyes converge or diverge to compensate for the effect of the prisms, the distant target is seen singly (though apparently at a nearer or farther distance), and the images of the slides remain superimposed. If however the eyes fail to compensate for the prisms, the images fall off the fovea of the non-fixing eye, and the slides are no longer seen superimposed.

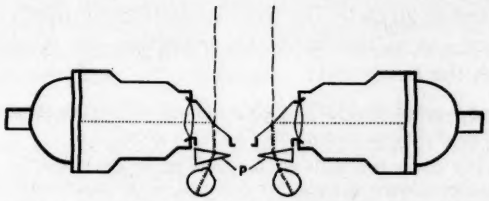

FIG. 4.—Principle of method used for measurement of fusion.

If the target used is the usual one with the overall pattern, the stimulus to fusion is both central and peripheral, and is the maximal one available for the patient; if central suppression is present, the effective stimulus will, of course, be peripheral only. If reproduceable measurements of fusion are required, the illumination of the target and the slides must be standardized.

(*d*) Interrupted reading can be carried out by using slides such as those shown in Fig. 5 (overleaf). In the "friend" test (Fig. 5*a*), the capitals on one of the slides are angled so as to discourage fusional movements which, with Roman capitals, may superimpose F on R, I on E, and N on D with consequent difficulty in interpretation.

(*e*) Stereoscopic vision can be tested in the usual way, using slides with a black background.

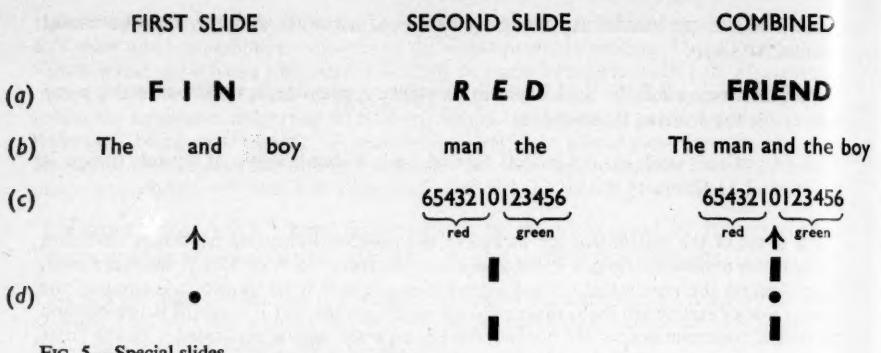

FIG. 5.—Special slides.
a, b.—Interrupted reading.
c.—Fixation disparity.
d.—Accurate measurement of subjective angle.
All slides are white on a black background, except *c*, in which the left-hand set of figures 654321 is red, and the right-hand set of figures 123456 is green.

By a suitable combination of these tests on the modified instrument, the binocular vision used under normal viewing conditions can be assessed. In most patients, of course, the findings confirm the impression given by the cover test and major amblyoscope readings; in such cases the modified instrument performs a similar function to say, Worth's four-dot test, with the advantage of being carried out under much more physiological conditions. When the impression given by the routine instrument differs from that given by the cover test, as in the following examples, the modified instrument usually confirms the cover test:

Case 11, a boy aged 6 years, had had a right accommodative convergent squint since the age of 4 years. Ten orthoptic treatments had been given.

Examination.—The cover test with glasses showed no deviation for near or distance; without glasses, the eyes were straight for distance, with occasional manifest squint for near. Routine major amblyoscope readings with glasses showed fusion at +10° with ductions from +5° to +20° only, but the modified instrument showed S.M.P. and fusion at 0°.

Case 12, a girl aged 7 years, had had a right accommodative convergent squint since the age of 4½ years.

Examination.—The cover test with glasses showed esophoria with rapid recovery for near and distance; without glasses, she had esophoria with rapid recovery for distance, but occasionally manifest squint for near. Routine major amblyoscope readings had always shown a convergent angle of +12° to +15°, with abduction only to +2°; on the modified instrument, she had S.P., fusion, and S.V. at 0°, and carried out interrupted reading without difficulty.

In some patients, however, additional information may be obtained, particularly those in whom the cover test is difficult to interpret:

Case 13, a girl aged 8 years, had had a right convergent comitant squint since the age of 4 years; it had appeared to be largely accommodative, and three short courses of orthoptic

treatment had been given at approximately yearly intervals since the age of 6 years, after which routine examination on the major amblyoscope showed S.M.P. at +6° with fusion from −2° to +15°, and stereopsis. Worth's four-dot test indicated binocular single vision. The child was thought on many occasions to be controlling the deviation well.

Examination.—When she was seen for assessment her eyes sometimes appeared quite straight for distance and near with glasses, but occasionally a minute flick was seen indicating right convergent squint; without glasses, she had a manifest convergent squint for near and distance. The cover test, however, was made difficult by a strong habit of nodding the head, which was particularly marked during the cover test and also when the glasses were removed; she was also very fidgety, and was being treated with bromide by her own doctor. Examination on the modified major amblyoscope with glasses showed S.M.P. at +4° (*i.e.* a convergent angle of 3° for testing at 3 metres), indicating that she was not, in fact, using bifoveal fixation under normal conditions. The measurements on the modified instrument were done quite easily and repeatedly.

Case 14, a girl aged 6 years, had had an alternating convergent squint since the age of 3 years with slight overaction of the right inferior oblique muscle but no definite muscle paresis. The angle of squint had always been greater for near than for distance, and she had at various times been thought to show no deviation for distance, a slight alternating deviation, or an esophoria. She had never needed occlusion, though the visual acuity in the right eye had tended on occasion to be slightly less than that in the left. The angle on the major amblyoscope had always been about 15°, with S.P. and fusion with weak ductions, but with a marked tendency to central suppression.

Examination.—When she was seen for assessment, the cover test with glasses for distance appeared to show esophoria of about 10° with a very slow recovery, but it was uncertain if the eye did in fact recover to the straight position, giving the impression of right convergent squint with a fixation disparity that was not fully established. Examination on the modified instrument with foveal slides showed an angle of +5° with a tendency to central suppression, showing that the condition was not an esophoria (in which case the reading would have been +1° at 3 metres).

Occasionally, both the routine major amblyoscope readings and the cover test may agree in indicating a good functional result, whereas the modified method indicates that this is not so:

Case 15, a girl aged 10 years, had had a left convergent accommodative squint since the age of 4 years.

Examination.—The visual acuity was 6/6 in the right eye and 6/6 partly in the left. The cover test with glasses showed no deviation for near or distance; without glasses, she showed esophoria with rapid recovery for near and distance. Routine major amblyoscope readings with glasses showed fusion at +2° with ductions from −8° to +15°, and some stereopsis; Worth's four-dot test with and without glasses indicated binocular single vision at all distances. On the modified instrument, however, she showed S.M.P. at −1° with glasses, and at −2° without glasses, at 3 metres, and definite alternation on attempting interrupted reading.

Further examples of small-angle squints, in which more detailed examination was carried out to elucidate the exact conditions present, are given below (Cases 16 and 17). In all these patients the reproduceability of the readings is noteworthy; if binocular vision (whether as bifoveal fixation, heterophoria, or fixation disparity) is being used under normal conditions, the fusion

produced by the distance fixation target maintains a steady position of the eyes relative to each other, so that readings with suitable slides are consistently reproduced to 0·5 prism dioptre.

SPECIAL INVESTIGATIONS

Peripheral Fusion.—This can be tested separately, even in the presence of central fusion, by covering the central part of the distant target by a matt grey square or circle of stiff paper or board (Fig. 6); the arms of the modified instrument are set so that the images of the slides are superimposed in the centre of this circle. The power of fusion is then measured as already described, the stimulus to fusion now being entirely paramacular or peripheral. Different sizes of circles can be used to eliminate greater or lesser

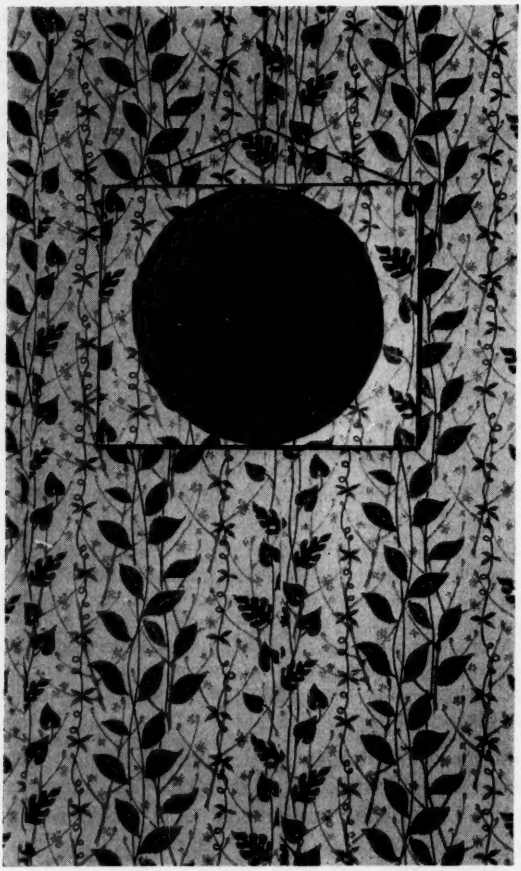

FIG. 6.—Distant target for measurement of peripheral fusion.

areas of central fusion. It is convenient to hang the circle by an adjustable cord from a hook placed at the top of the target, so that its height can be altered to correspond with the most convenient position of the instrument for the particular patient being tested.

The effectiveness of the method is well illustrated by a patient with visual acuity of 6/36 in each eye due to myopia; the corrected visual acuity was 6/6 in each eye; peripheral fusion, measured with a central grey area 10° in diameter, produced adduction of 8△ without glasses and 14△ with glasses; with prisms just stronger than these, so that the images of the slides were seen separated, removal of the central grey area provided immediate superimposition, due to the extra ductions produced by central fusional stimuli. In most people, however, peripheral fusion appears to be approximately as strong as that produced by the whole target.

Fixation Disparity.—To measure significant amounts of fixation disparity, slides more accurate than the usual S.M.P. slides are required. Those shown in Fig. 5(*c*) are convenient; if the arms of the instrument are locked in the position corresponding to no deviation and bifoveal fixation is perfect, the arrow points to the figure 0, but if fixation disparity is present the arrow will point to one of the figures to the right or left of 0, depending on the direction of the fixation disparity. It is convenient to colour the figures on one side red, and on the other side green, and to space them so that they give a direct reading of the amount of fixation disparity in prism dioptres or degrees. The principle of this method is fundamentally the same as that of the Maddox rod and tangent scale, or Maddox wing; with the arms of the instrument locked at 0° for distance fixation, the images of the arrow and of the figure 0 fall on the fixation points of the two eyes (Fig. 7); in fixation disparity, one of these points will be non-foveal. If the numbers are seen by the eye with non-foveal fixation (Fig. 7*a*) the fovea F will receive the image of one of the numbers other than 0; since the retinal correspondence in true fixation disparity is normal (all subjective methods of measuring fixation disparity depend on

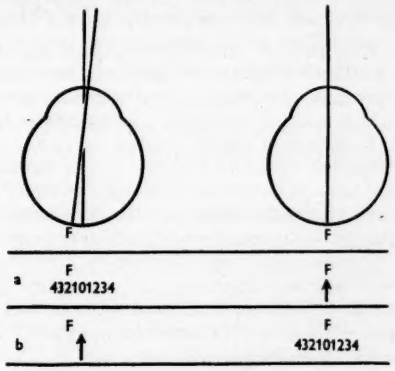

FIG. 7.—Principle of slides for measurement of fixation disparity, showing relative positions of fovea and image of slide in each eye, in a patient with left fixation disparity of 2△
(*a*) Arrow opposite eye with foveal fixation.
(*b*) Arrow opposite eye with non-foveal fixation.

this), the arrow, seen foveally by the other eye, will therefore be seen to lie opposite this number. Similarly, if the arrow is seen by the eye with non-foveal fixation (Fig. 7*b*), its image will fall on a non-foveal point and will be seen to lie opposite the number which lies on the corresponding non-foveal point on the retina of the other eye. It is immaterial, therefore, which eye sees the arrow and which one the number (neglecting aniseikonia, which is of no significant magnitude in this connexion).

An alternative method of measuring fixation disparity depends on the accurate determination of the subjective angle, since, in the presence of normal correspondence, this gives directly the angle between the two foveal visual directions. For this purpose, ordinary S.P. or S.M.P. slides are not sufficiently accurate, and are better replaced by slides depending on vernier alignment (Fig. 5*d*) analogous to those described by Jampolsky (1951). Such measurements can be made with remarkable accuracy and reproduceability by patients with fixation disparity.

The presence of fixation disparity may be easier to demonstrate if the central area of the distant target is occluded by the grey circle as in measurements of peripheral fusion, since it has been shown (Jampolsky, Flom, and Freid, 1957) that absence of central details in the binocular target allows greater fixation disparity to occur; most measurements of fixation disparity (such as those of Ogle, 1950) have indeed used a small central blacked-out area.

All these measurements require precise centring of the slides, which must not wobble in the holders, and accurate setting for the interpupillary distance; the adjustment of the arms of the instrument should be checked subjectively by the operator to ensure that it corresponds to the position of no deviation for the particular target distance used.

The value of these measurements in the accurate diagnosis of small-angle squints is shown by the following patients:

Case 16, a man aged 42 years, was first seen in the orthoptic department at the age of 40 years with a history of left convergent squint for 4 years only, following a blow on the left eyebrow, and intermittent diplopia which was increasing in frequency and severity.

Examination.—He had a negligible degree of hypermetropic astigmatism, the corrected visual acuity being 6/6 in each eye; the cover test showed a constant left convergent squint for near and distance; the ocular movements were full; the major amblyoscope showed S.M.P. and fusion at +32°, with ductions from 30° to 35°, and doubtful stereopsis.

Recession of the left medial rectus and resection of the left lateral rectus were performed at the age of 42 years, and he was seen for assessment 6 months later. The cover test showed esophoria with rapid recovery to a position which often appeared straight, but which occasionally gave the impression of very slight fixation disparity of the left eye. He had slight limitation of adduction in the left eye.

Routine major amblyoscope readings showed S.M.P. (unsteady) at +5°, and fusion at +4° with adduction to +20° and abduction to 0° with a tendency to left suppression; weak stereoscopic vision was present. On the modified instrument, the subjective angle was +1·5 to +2° (a convergent angle of 0·5 to 1° at 3 metres) depending on the relative levels of illumination of the target and the slides; using the fixation disparity slides, he

showed disparity with a convergent angle of 0·5 to 1°. When the target illumination was high, this could only be elicited with the grey circle over the central area of the target, showing the dependence of the magnitude of the fixation disparity on the conditions present.

Case 17, a boy aged 6 years, had had a right convergent comitant squint since the age of 18 months, with only a slight hypermetropic astigmatism, which was not worth correcting.

Examination.—The visual acuity was 6/12 in the right eye and 6/6 in the left. The cover test showed a slight but definite manifest right convergent squint for near and distance with no recovery.

Routine major amblyoscope readings showed S.M.P. at 0° with fusion with ductions from −5° to +12° and S.V., but there was definite movement of the right eye on flashing when fixing with the left eye; it was thought that fixation disparity might be present. Examination on the modified instrument showed an objective angle of +3°, but a subjective angle of +1° (*i.e.* harmonious A.R.C. for examination at 3 metres). The fixation disparity slides showed no fixation disparity. There was no suppression on either the routine or the modified instruments. Peripheral fusion was almost as strong as central fusion, abduction being 8△ with the complete target and 6△ with a central occluded area of 10°.

The condition appeared to be a small degree of esotropia (2° at 3 metres) with abnormal retinal correspondence of the type described by Jampolsky (1951), though it is doubtful if this type of A.R.C. is of the same nature as that seen in the large-angle squints.

Discussion

The modified method of examination obviously is not intended to replace accurate cover testing and examination on the routine major amblyoscope as the basis of assessment of patients with squint. It does, however, provide some additional information; it is not intended in the present paper to assess its value fully, since this can only be done over a long period of time, but several points of interest emerge from this preliminary survey.

(1) In those patients in whom the modified instrument confirms in general the findings of the cover test and routine major amblyoscope readings, but in whom it indicates an appreciably different angle from that found on the major amblyoscope, the significance of the measurements in relation to the surgery to be performed can only be assessed over a long period. The readings on both the modified and routine instruments have to be taken into account, those with the modified instrument indicating the normal position of the eyes, and those with the major amblyoscope the fully dissociated position. It is possible that the readings with the modified instrument may help to elucidate cases in which surgery gives an unexpectedly large or unexpectedly small change in the apparent angle of squint.

(2) The information obtained from the modified instrument regarding the state of the retinal correspondences may be of considerable importance. It is obviously of importance to know as soon as possible when abnormal retinal correspondence is present, and the modified instrument may reveal this condition long before it is detected on the routine major amblyoscope (*e.g.* Case 8); in such patients, as Halldén (1952) has suggested, the presence of

harmonious A.R.C. before operation may be responsible for a tendency to revert after operation to the original angle of squint. On the other hand, the presence of A.R.C. which is harmonious under normal viewing conditions may be of value in stabilizing the position of the eyes and preventing subsequent divergence. Further experience will also show whether or not the presence of A.R.C. which is detectable only by the modified method has the same gloomy prognosis for the subsequent development of normal binocular vision as has A.R.C. on the routine major amblyoscope.

The findings on the modified instrument are also of considerable interest regarding the aetiology of this condition; the fact that the A.R.C. in normal viewing conditions is commonly harmonious lends support to the theory that it is a purposive reaction, and the instrument lends itself to the investigation of purposive fusional movements in such patients in ordinary clinical practice without the use of projectors such as were used by Halldén (1952). It is not surprising that A.R.C. is more easily and consistently demonstrated on the modified instrument than on the routine major amblyoscope, since the distant target forms a strong stimulus to purposive adaptive phenomena.

(3) Since the instrument measures the binocular use of the eyes under normal viewing conditions, it has obvious advantages over the Worth four-dot test and similar devices. It may help to elucidate those patients who, in spite of an apparently excellent result as judged on cover test and routine major amblyoscope readings, have poor depth perception (Naylor, Shannon, and Stanworth, 1956); Case 15 may well come into this category.

(4) The measurement of peripheral fusion (in small-angle squints) is brought within the range of clinical practice without the use of complicated apparatus unfamiliar to the orthoptist, such as that used by Burian (1941) and Winkelman (1951). The patient also finds these measurements much easier than by the use of special slides in the routine major amblyoscope, such as were used by Swan and Laughlin (1944) and Stanworth (1952, 1955). The measurement of peripheral fusion may be of particular interest (Stanworth, 1949; Cashell, 1950) in those patients who, having no appreciable binocular vision before operation, somewhat unexpectedly develop binocular vision after operation, as assessed on the major amblyoscope; the stages by which this result is achieved cannot be elucidated by the cover test and routine amblyoscope readings alone, but the modified instrument may permit investigation of the suggestion (Stanworth, 1952) that peripheral fusion is an important factor.

(5) The measurement of fixation disparity is also brought within the range of clinical practice. The method is analogous to that used by Ogle (1950), but is not, of course, so accurate, and will not measure the minute amounts of fixation disparity (in fractions of a degree) found in heterophoria. It is designed to measure degrees of fixation disparity which may be of clinical importance; the cover test will only definitely diagnose those patients who have fixation disparity combined with a greater amount of heterophoria, so

that the affected eye shows partial recovery when uncovered; it appears probable, however, that fixation disparity may occur without extra heterophoria, and so would not be detectable by the cover test. The instrument will thus facilitate the diagnosis of small-angle squints, as in Cases 16 and 17, and help to elucidate the frequency of fixation disparity and anomalous correspondence in such patients.

SUMMARY

A modified major amblyoscope is described in which measurements are made in a similar manner to those with the routine instrument, but in which the conditions for the patient approximate to those of everyday life. The changeover from routine to modified method of examination is rapid and easy.

It makes possible an assessment of the angle of squint and the use made of the patient's binocular vision under normal conditions. It is also of particular value in the detection of abnormal retinal correspondence, in small-angle squints, and in patients in whom the cover test is difficult to interpret. The measurement of peripheral fusion (in small-angle squints) and of fixation disparity in normal viewing conditions is brought within the range of ordinary clinical practice.

I am greatly indebted to Dr. E. J. Naylor for his collaboration in the early stages of the development of this instrument; to Miss E. Stringer and the staff of the Orthoptic Department of Manchester Royal Eye Hospital, especially Miss D. da Cunha and Miss M. Slater; to Mr. A. B. Nutt for his comments and encouragement; and to the Medical Illustration Department of the Manchester Royal Infirmary for Figs 3, 4, 6, and 7.

The instrument is manufactured by Curry and Paxton Ltd., who gave great help in the detailed design.

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A CONTRIBUTION TO THE EXPERIMENTAL STUDY OF THE ALLERGIC PROCESSES OF THE ANTERIOR SEGMENT OF THE EYE*

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In order to observe local anaphylactic processes caused by the injection of antigenic substances into the anterior segment of the eye, a series of experiments on 108 rabbits was undertaken, using human, horse, pig, and ox sera as antigens. Each series consisted of six to twelve animals, and included control groups when needed.

Procedure

The injections were made with the animals under general anaesthesia (about 40 mg. "Nesdonal" per kg. body weight).

(1) With the eye rotated downwards by pulling the superior rectus muscle, the needle was inserted obliquely into the central area of the cornea and 0.1 ml. antigen was injected.

(2) The needle was inserted obliquely through the conjunctiva about 2 mm. from the limbus until it appeared in the anterior chamber; the aqueous was then allowed to drain off, and 0.1 ml. antigen was injected.

Results

These are given in Table I; 28 animals underwent a double sensitization of both cornea and uvea in the same eye with the same antigen. The results are analysed by site in Table II, and the antigens used are shown in Table III.

The injection of antigen into the eye was immediately followed by a traumatic reaction (oedema of the cornea, iris congestion, and sometimes a little exudation) which disappeared completely within a few hours. Control animals, in which one eye was sensitized and the other was injected with a 1 per thousand dilution of "Sunoxol", a preservative used in the sera, showed a similar traumatic reaction in both eyes, which was due to the fluid introduced. The eyes which were given "Sunoxol" only showed no other reaction either primary or secondary.

Some of the results are illustrated graphically in Figs 1 to 3 (overleaf), which show the height and duration of the reactions.

The "height" of the corneal reaction was judged arbitrarily in degrees of transparency:

(*i*) Slight cloudiness;
(*ii*) Intermediate;
(*iii*) Total opacity.

* Received for publication May 7, 1957.

TABLE I

ANALYSIS OF RESULTS

Sensitization		Monocular		Binocular		Remarks
		Cornea	Iris	Cornea	Iris	
Reaction	None	21	6	13	6	
	Primary Early	16	15	21†	9	† Seven animals reacted monocularly
	Primary Late	8	0	1	0	
	Primary Early + Primary Late	2	0	3	0	
	Primary Early Prolonged + Primary Late ..	1	2	9	3	
Second Dose Administered (intravenously)		21	20	23	19	
Second Response Obtained ..		7	5	12*	13	* *Four animals showed* only a contralateral reaction

TABLE II

RESULTS ANALYSED ACCORDING TO SITES OF REACTIONS IN THE SAME EYE

Reaction	Primary	Present	Present	Absent	Absent
	Secondary	Present	Absent	Present	Absent
Site	*Cornea*	*15*	*12*	*4*	*13*
	Iris	16	11	2	10

TABLE III

RESULTS ANALYSED ACCORDING TO SITES OF REACTIONS TO DIFFERENT SERA

Serum Used as Antigen	Site		Primary Response		Second Dose Administered		Second Response Obtained	
	Cornea	Iris	Cornea	Iris	Cornea	Iris	Cornea	Iris
Human	37	12	25	12	19	9	12	3
Horse	22	13	12	7	14	8	8	4
Ox	65	35	48	25	24	21	6	10
Pig	7		5		7		2	
Total	131	60						

The "height" of the reaction in the anterior uvea was judged as follows:

(*i*) Congestion of the iris, miosis, and a little exudation;

(*ii*) Iris oedema and haemorrhages with exudation;

(*iii*) Marked exudation with synechiae.

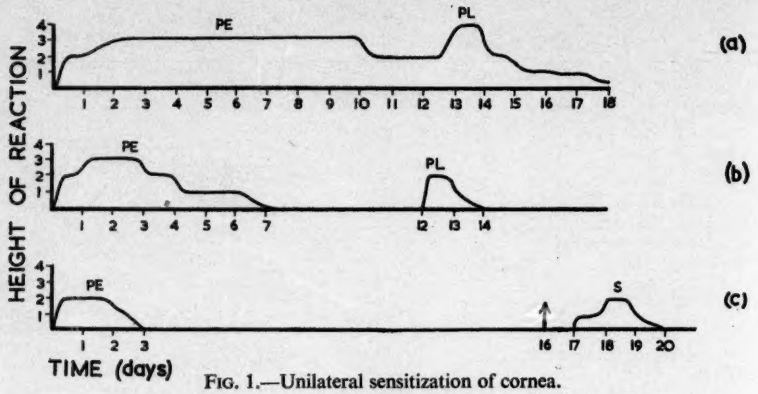

FIG. 1.—Unilateral sensitization of cornea.
PE=Primary early. PL=Primary late. S=Secondary.

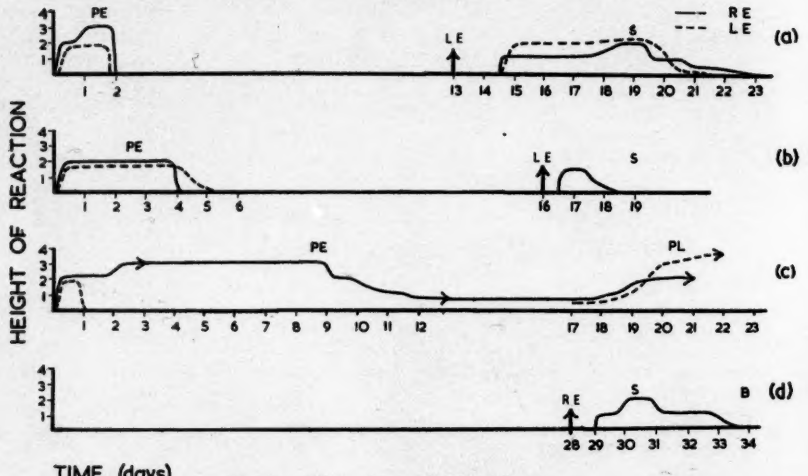

FIG. 2.—Bilateral sensitization of cornea.
PE=Primary early. PL=Primary late. S=Secondary.

About sixty animals were given a different antigen into each eye as the primary dose, and in these cases the results are represented on the graphs by full and dotted lines.

The prolonged type of primary early reaction (Figs 1*a*, 3*b*, and 3*c*) was seen in fifteen instances, and was followed by a late reaction in each case. In bilaterally-sensitized animals, the primary late reaction, when it appeared, occurred in both eyes.

Second Doses.—In each Figure, arrows indicate the intravenous administration of a second dose of 2–3 ml. antigen, and above the arrows is shown the eye which had already been sensitized to the same antigen.

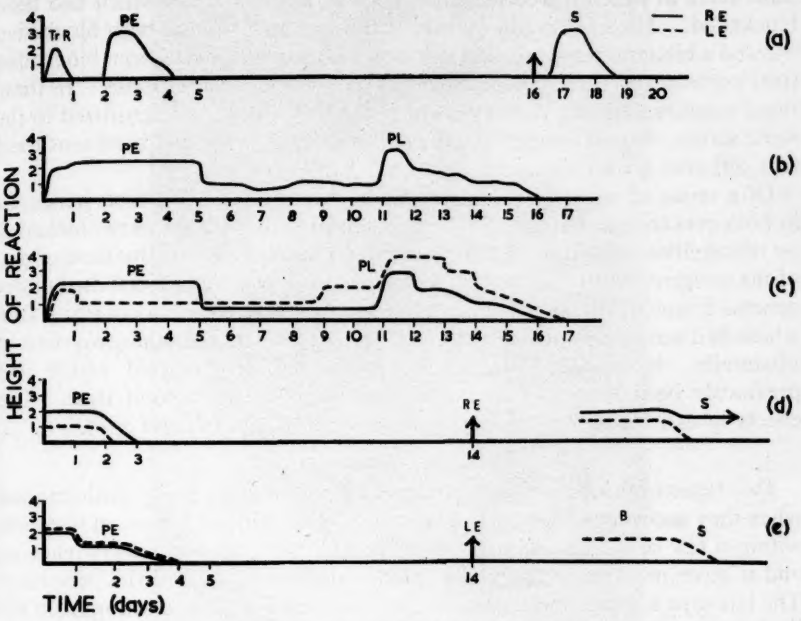

FIG. 3.—Unilateral and bilateral sensitization of iris.
PE=Primary early. PL=Primary late. S=Secondary.

The binocularly-sensitized animals were given as a second dose only one of the antigens previously used. In all cases which reacted to the second dose, a contralateral, non-specific response was also obtained. Four animals in which the two corneae had been sensitized to different sera, showed *only* a contralateral response, the eye which had already been sensitized to the antigen used for the second dose remaining quiet. The control animals, which were given as a second dose a serum other than those used for the first sensitizing injection, did not react at all (these included eight animals which had been given human or ox serum, sheep serum being used for the "anaphylactic" dose).

Effects of Blockading the Eyes.—A series of nine animals which had been binocularly sensitized in both the cornea and the uvea (the right eye with human serum, and the left with ox serum) received a second dose (of human serum only) on the 28th day after the first injection. Just before the second injection, four of them received bilaterally a "blockading" retrobulbar injection of 2 ml. absolute alcohol and 4 per cent. Novocain in equal amounts. The corneae of only three animals reacted: two which had been blockaded and one which had not. The uveae of four animals reacted: the

same three in which the corneae had reacted, and one other which had been blockaded. The one reactor (cornea and iris) which had not been blockaded showed a bilateral response, and the three reactors which had been blockaded (two corneae and three irides) showed only a monocular response. In these three reactors, only the right eye, which had previously been sensitized to the same serum, reacted, while the left eye, which had previously been sensitized to a different serum, remained quiet.

Of a series of six animals, in which the anterior uvea had been sensitized in both eyes (one to human and the other to ox serum), three were blockaded by retrobulbar injections, and all were given a second intravenous dose of one of the antigens used in the first injections. One blockaded animal died under general anaphylactic shock, and the other two reacted monocularly. One which had not been blockaded did not react at all, and the other two reacted bilaterally. Here again, only those blockaded eyes reacted which had previously been sensitized to the antigen used for the second dose. The effects in two blockaded cases are illustrated in Figs 2 (*d*) and 3 (*e*).

Discussion

Two types of primary response may occur. These are easily distinguished when they occur in the cornea. The early type appears at the site of injection within a few days, spreads toward the limbus but does not always reach it, and is more marked centrally, its intensity decreasing toward the periphery. The late type appears some 10 days after the local injection, starting from the limbal region in the form of a whitish-grey ring, and progressing to the centre of the cornea; a dense network of blood vessels arranged concentrically like a wreath arises from the limbal plexus (Fig 4, opposite). Both types of response may occur successively, or one or both may fail to appear.

The secondary response, when elicited, has the same clinical features as the primary late response.

The following facts are to be noted:

(1) A primary early reaction, when marked and prolonged, was always followed by a late one.

(2) In binocularly sensitized animals, the primary late reaction, when it appeared, was bilateral.

(3) Nearly all binocularly sensitized animals which reacted to the second (intravenous) dose, did so bilaterally, although only one of the original antigens was re-introduced.

(4) Sometimes the processes appeared in each eye successively. The specific secondary responses were the first to appear.

These observations suggest the existence of a relationship between the parallel processes in the two eyes, and indicate that a process in one eye can set going or contribute towards a similar process in the other eye. The secondary specific process may result from an interaction between some residual antigen stored in the cornea and the circulating antibodies, the production of which has been stimulated by the second dose. It may also result from

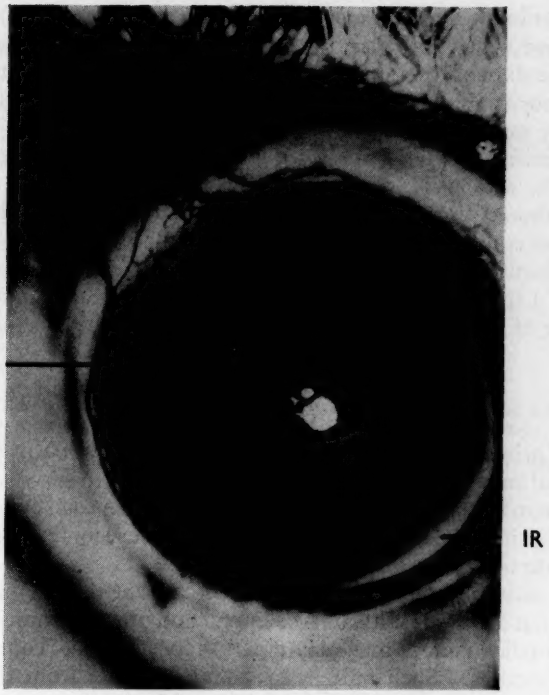

FIG. 4.—Primary late reaction of cornea.
VR = Vascular ring.
IR = Infiltration ring.

direct contact between the re-injected antigen and the reactive agents already stored in the corneal tissue. This latter course is the more likely, as it does not seem very probable that a residual antigen will persist in the corneal tissue, especially after repeated secondary responses. The contralateral, non-specific effect could also perhaps be explained by an analogous humoral mechanism, the production of antibodies being also stimulated in the fellow eye (by an anamnestic reaction).

This explanation seems insufficient, however, to account for the results in the blockaded cases; the specific response could not be influenced by the retrobulbar injection, and the contralateral response was invariably prevented by it. The blockade could not have influenced the humoral relations of the two eyes, for the second dose of antigen had the same chance of reaching both eyes as the antibodies the production of which had been stimulated by the second dose.

The blockade may, however, influence the neuro-reflex relationship between the two eyes, and the results obtained in these experiments indicate the existence of a reflex factor in the allergic processes of the eye.

In four animals a contralateral effect was obtained only after the second dose; the eye which had already been sensitized to the antigen used for the second dose did not react, but its fellow did (Fig. 2*b*). This may be due to an "elective sensitization" of tissues (Føss, 1949). The absence of response in the first eye might be due to a decreased level of concentration of the corresponding antibodies. The supposed increased in the antibodies in the fellow eye might be responsible for its non-specific response. If this explanation is correct, it should also be applicable to the cases which reacted bilaterally, but in these the contralateral reaction was prevented by the blockade. If the reflex mechanism does play a part in this contralateral response, even in the absence of a specific response in the fellow eye, one must admit the existence of a source of the reflexes which seem to be responsible for this non-specific reaction.

Conclusions

(1) The primary early reaction in sensitized eyes, when marked and prolonged, is always followed by a late reaction.

(2) In animals sensitized with a different antigen in each eye, a marked and prolonged primary early reaction, even in one eye only, is followed by a bilateral late reaction.

(3) The results of these experiments suggest that the allergic processes of the eye do not always result from the interaction of the humoral elements, but that neuro-reflex factors may play a part in the genesis of contralateral, non-specific responses. Such reflexes probably originate from the pre-existing process in the fellow eye. In principle, this idea may be extended to cover analogous reactions between the eyes and any focal process elsewhere, or even between any two organs.

(4) The occurrence, though rare, of a contralateral, non-specific reaction in the absence of a specific reaction in the fellow eye, suggests that even an apparently normal unaffected eye may evoke pathogenic impulses in the other eye.

These experiments were mainly carried out at the Ophthalmic Clinic of the Medical Faculty at Nantes, France, in 1956. The Principal at the time, the late Prof. G. P. Sourdille, arranged for my work to be carried out under the most favourable conditions. He and his successor, Prof. J. Legrand took a great interest in my work, giving me their best advice. I should like to express my most cordial appreciation for all this help, and also that of Dr. Hervouët, head of the Clinic laboratory, who assisted me personally and ensured the availability of all the facilities of the laboratory, and of M. A. Lenoir, who gave technical help and was responsible for the photography and histopathological preparations.

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STURGE-WEBER'S SYNDROME*

REPORT OF AN UNUSUAL CASE

BY

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No case exhibiting so many interrelated abnormalities as were seen in this patient could be found in the available literature.

Case Report

A man aged 26 was admitted to hospital on October 21, 1954, complaining of failing vision in the right eye. No relevant family history could be obtained and he had had no serious disease before the age of 10 years, when he became ill with fever and skin eruption. While in Persia in 1941, he had been treated for trachoma in both eyes, and at that time the visual acuity of the left eye was so poor that he was unable to read with it. In 1950, pain and aching all over the body were treated by his own doctor, to whom he gave a history of frequent epistaxis since childhood. About Easter, 1951, the visual acuity of the right eye began to deteriorate and he was compelled to give up driving lessons and to leave his work.

On admission to hospital, besides impairment of visual acuity, he also complained of transient slight headache, confined mainly to the occipital region and the back of the neck, which he noticed especially when bending down and after effort.

Examination.—He appeared to be well-built and well-nourished. The lungs, heart, and abdomen showed no gross abnormality. The face was covered with flat patches of naevus flammeus, corresponding roughly with the trigeminal area. Similar patches were seen on the soft and hard palate (which was unusually high), and on the arms and the left side of the chest. The vascular changes were much more pronounced on the left side of the body, especially on the left arm (Fig. 1, opposite). Among the vascular spots there were also whitish areas of depigmentation of the skin, clearly seen on the left side of the chest and the left arm. The anterior incisors were better developed than the lateral ones, and the canines were pointed.

Both eyeballs, especially the left one, showed some degree of proptosis. On both sclerae there were extensive patches of slate-grey pigmentation. Brown flecks were seen at the openings of the canals of the ciliary vessels. The corneae were clear, Descemet's membrane intact, and the anterior chambers quite deep, the left one apparently the deeper.

Both irides were dark brown and showed in their lower parts a marked division by a well-developed iris frill (collarette), which reached the pupil in numerous and dense radial ridges. The upper part of the iris frill seemed to be non-existent, but a more detailed examination showed that it was only less pronounced and displaced towards the pupils, which made the pupillary zone of the iris much narrower. The sphincter design was missing in the upper parts of the irides.

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FIG. 1.—Showing distribution of naevus patches, more pronounced on left side of face and trunk, slight proptosis of left eye, and depigmented areas underneath left nipple and on left arm.

The pupillary margin was of normal shape when seen from below, but from above it appeared as a smooth dark-brown strip without crenations. The pupils were pear-shaped with the stalk in the left eye at about 11 o'clock (Fig. 2), and in the right eye at about 1 o'clock.

FIG. 2.—Pear-shaped pupil of left eye.

Their reaction both to light and to accommodation was very sluggish and confined to the area of the normal iris. The media were clear and transparent.

In the right fundus there was pronounced tumour-like papilloedema of long-standing and about 7D elevation. On the temporal side and above the swollen optic disc, there was a small tortuous angioma-like vessel, terminating in an old haemorrhage. The visual acuity was 6/60. The ocular tension was normal.

In the left fundus there was pronounced cupping (about 5D) of the disc, with advanced optic atrophy. The ocular tension was 40 mm. Hg (Schiötz). The visual acuity was almost 6/60. Transillumination of the eyeballs failed to reveal any abnormal shadows.

Laboratory Investigations.—Urine normal. Blood count normal. *X*-ray of skull, no major abnormality. No other abnormalities in the central nervous system.

Diagnosis.—The presence of advanced papilloedema suggested the probable existence of a space-occupying cerebral lesion, and, on the advice of Dr. E. R. Bickerstaff, the patient was transferred to the Midland Centre of Neurosurgery. A bilateral arteriogram showed a large temporal tumour.

Operation.—Craniotomy was performed on December 1, 1954, and a very large tumour was discovered in the right temporal lobe, rising from the lateral fissure and surrounded by an area of haemorrhage. Though its appearance was rather odd, it was thought from the way it shelled out to be a meningioma. Section of the tumour, however, showed it to be a melanoma, which was thought to originate in the eye.

Pathological Report on the Tumour and Cyst Walls: "The specimen consists of a large number of pieces of friable, haemorrhagic tumour. The section shows a typical melanoma, some areas of which are completely amelanotic. No mitoses are to be seen".

Progress.—At first the patient's post-operative condition was good. No abnormal neurological symptoms were found, and gradually the papilloedema of the right eye receded (Fig. 3).

The visual acuity in the right eye deteriorated but slowly recovered. After the initial period, however, the patient showed marked mental disturbance; he later improved considerably but never regained his pre-operative mental condition.

Some weeks after the operation, the papilloedema had subsided and the angioma-like vessel and haemorrhage had disappeared, but the patient began to deteriorate (Fig. 4).

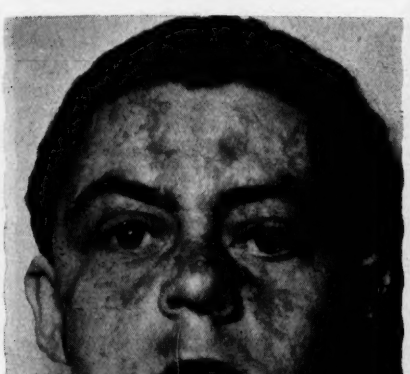

FIG. 3.—Shortly after operation.

FIG. 4.—Showing relapse of brain tumour, butterfly-like distribution of facial naevus, slight proptosis of left eye, and underdeveloped second incisors.

He first developed swelling of the liver, and then swelling of the skull flap; he became dull and confused, and finally a left hemiparesis appeared, affecting the arm more than

the leg. It was apparent that these symptoms were due to local relapse and to secondary deposits. The patient grew rapidly worse, lost control of sphincters, became semi-conscious, and died on July 27, 1955. Permission for a *post-mortem* examination was not granted.

Discussion

Melanosis of the Sclera.—According to Duke-Elder (1942a), this is not a rare condition. It may progress rapidly in youth and more slowly in age, but often remains more or less stationary. The possibility of a more serious prognosis has to be borne in mind, and the high incidence of malignant change makes it necessary to regard these cases with some anxiety.

Pear-Shaped Pupils.—These are described as extremely rare by Duke-Elder (1942b), who includes them with "pupil anomalies other than colobomata".

A few cases similar to that described above were reported by Liebitzky (1929), who stated that the main symptoms of this iris anomaly were pear-shaped pupils, simplified structure of the pupillary margin, and local absence of the sphincter.

Whatever the cause of this anomaly, the present case seems to prove that pear-shaped pupils cannot be regarded as a mere variation of an atypical coloboma. The symmetrical structure of the pupils in this case may be due rather to a primary inhibition in development of a sector of the optic cup than to accidental inhibitory effect of a persistent capsulo-pupillary vessel.

Sturge-Weber's Syndrome.—Sturge (1879) described this disease as "congenital glaucoma with coexistent naevus flammeus and epileptiform fits of the opposite side of the body". He postulated a focal lesion of the cerebrum similar to that found on the face. In Sturge's patient, one pupil became oval in shape with the narrow end downwards after homatropine. Parkes Weber (1929) reported a patient with right-sided hemiatrophy, right-sided congenital spastic hemiplegia, widespread cutaneous vascular naevi with bilateral involvement of the face, and left glaucoma. Skull *x* rays revealed that the left side of the brain was more opaque than the right. No epileptic fits occurred. Weber later succeeded in obtaining radiological evidence of intracranial calcifications, which he regarded as tortuous and calcified blood vessels.

Cerebral and Neural Involvement.—Krabbe (1934, 1955), having performed a biopsy on one of his cases, was able to demonstrate that the calcifications were not confined to the pial vessels but also involved the cerebral cortex. He found angiomatous changes of the pia and focal aplasia of the brain, with probable secondary sclerosis and calcifications of the aplastic area. This developmental defect was thought to be both ectodermal and mesodermal in origin. He suggested that Sturge-Weber's syndrome originated in early foetal life many weeks before the development of the trigeminal nerve, and that the distribution of the naevus vasculosus in the

trigeminal area was merely accidental. The abortive forms could be considered only on the understanding that the disease had no distinct limits. The term Sturge-Weber syndrome should be applied only in cases showing both the naevus on the face and intracranial calcifications of the special type demonstrable on *x*-ray films.

Kautzky (1949) suggested that the Sturge-Weber syndrome was due to an inborn inhibition of the parasympathetic part of the cerebrospinal nerves, mainly the trigeminal nerve.

Van Bogaert (1950) described the Sturge-Weber syndrome as essentially a cutaneous and meningeal angiomatosis. Although cerebral angiomatosis was found in many cases, the cortical changes were secondary to the mesodermal defect, thus showing an atrophy, not aplasia. He drew attention to the relation between pigment dystrophies and angiomatoses and presumed the existence of neurodermal melanosis although he was unable to find a case wherein the ocular melanosis was associated with meningeal melanosis in the literature.

Calcifications.—According to Livingstone, Eisner, Brown, and Boks (1956), the syndrome has three main features: a vascular naevus, mainly in the trigeminal area, radiological evidence of calcifications, and convulsions. The last appears to be a constant symptom, but the naevus and intracranial calcifications are not always present. In one case, reported by these authors, many *x*-ray examinations of the skull were made on numerous occasions between the ages of 3 and 18 years, but no evidence of calcification was seen.

It is not yet known how the mineral deposits of iron and calcium occur in the angiomatous and atrophic cortex. Some authors have underlined the role of lipoid breakdown and liberation of the phosphoric acid in preparing necrotic tissue for calcification, but much further investigation is necessary to solve this problem. Tingey (1956) reported the results of the chemical analysis of iron and calcium in the brain in three cases of Sturge-Weber's syndrome. A great deal of calcium was present in the white matter as well as in the cortex, but no excess of iron was found.

Frigyer, Mattyus, and Molnar (1953) stated that calcification was the most frequent symptom of the Sturge-Weber syndrome, but was not essential to the diagnosis. The vascular anomaly was the primary change and this explained the circumscribed nature of the clinical picture. The metabolic changes due to the vascular anomaly caused colloid formation, calcification, and destruction of the brain substance. The vessel wall was the primary site of deposition.

Heredity.—According to Bergstrand, Olivecrona, and Tönnis (1936), the Sturge-Weber syndrome was due to a congenital tendency which might be inherited, but there was not such clear evidence of heredity as in von Reck-

linghausen's and von Hippel-Lindau's diseases. The anatomical changes appeared immediately after birth, but grew larger as time went on, the brain and eye changes sometimes appearing later in life.

Related Conditions.—There is a close relationship between the Sturge-Weber syndrome and three other diseases involving vascular changes: those of von Recklinghausen, von Hippel-Lindau, and Bourneville. In von Hippel-Lindau's disease there are angio-reticulomata in the cerebellum, spinal cord, and retina; in Bourneville's disease there are angio-gliomata, renal angiomata, and angioma sebaceum Pringle on the skin; in von Recklinghausen's disease naevi anaemici appear on the skin; in the Sturge-Weber syndrome teleangiectases appear in the cerebellar cortex and choroid, angioma racemosum in the pia, and naevus flammeus on the skin.

Bergstrand and others (1936) also thought that a great number of cases of naevus flammeus over one or more trigeminal branches without any other symptoms were abortive forms of the Sturge-Weber syndrome. The symptoms in less severe cases might interchange in various ways. There was a frequent incidence of naevus flammeus and glaucoma or naevus flammeus with cerebral changes and epileptic fits.

Yakovlev and Guthrie (1931) regarded the vascular malformation as merely the most pronounced symptom of malformation of the brain, the developmental embryonic period at which the malformation began being alone responsible for the development of angiomatosis, neurofibromatosis, or tuberous sclerosis.

All these diseases belong to the so-called phacomatoses (φακωμα = birth mark) or hamartoses (ἁμαρτία = deviation, defect). Cases have been published in which these symptoms appeared together in one patient and were accompanied by other pathological changes. Bergstrand and others (1936) showed the Sturge-Weber syndrome to be associated with meningioma in one case, and with acromegaly in another.

Neural debility, obesity, sexual infantilism, highly arched palate, abnormally developed ears, flattened nasal base, and malformed teeth (better developed anterior incisors, undeveloped lateral incisors, pointed canines) have been described, as well as lesions more frequently found in tuberous scleroma and neurofibromatosis, such as cysts, congenital tumours of the viscera, etc.

Nonnenmacher (1955) published a report of a patient with Sturge-Weber's syndrome who simultaneously exhibited symptoms of Klippel-Trenaunay's (Parkes Weber) disease. A flat naevus vasculosus was scattered all over the body, especially on the face and arms, and more on the right than on the left side, capillary ectases appeared on the soft and hard palate, and there was bilateral buphthalmos, but calcifications were not found, and there were no epileptic fits. The flat naevus vasculosus and osteohypertrophy of the right side of the face and the right leg are typical of the Klippel-Trenaunay

syndrome. The same author included the neurodermal melanosis of Touraine (1949) among the phacomatoses.

Epileptic Fits.—According to Parnitzke (1956), the complete picture of the Sturge-Weber syndrome is characterized by facial naevus flammeus, congenital or acquired glaucoma, local or general convulsions, and calcified cerebral angiomata. Classic cases with fully-developed symptoms are infrequent but many authors state that at least two symptoms must be present to establish a diagnosis of Sturge-Weber disease. Parnitzke described four cases ranging from the fully-developed picture to a single symptom. The most interesting is his Case 4 in which only the typical, symmetrical intracranial calcifications were found; naevus, glaucoma, epileptic fits, and dementia were not present, but the patient had attacks of migraine.

Epileptic attacks occur only in patients with an inherent predisposition for convulsions. In some cases the calcifications are undetectable by radiological examination and can be found only by microscopy. Epileptic fits occur in the majority of cases, but Parnitzke's Case 4 seems to prove that the statistical data are artificial, because usually it is only the patients with epileptic fits who come to our notice. In such cases the epileptic fits appear much later in life and other abnormal neurological symptoms are absent.

Glaucoma.—The most important ocular complication of the Sturge-Weber syndrome is congenital or acquired early glaucoma, which does not necessarily occur on the same side as the naevus flammeus. Histological examination reveals angiomatous changes in the choroid. Various authors have reported optic atrophy; strabismus; angiomatous changes of the conjunctiva, sclera, iris, ciliary body, and retina; retinal glioma; choroidal atrophy; strands of persistent pupillary membrane; heterochromia of the irides; and exophthalmos. Inigo (1952) added posterior capsular cataract, and described a case which showed widespread angiomata of the conjunctiva and sclera with enlarged and tortuous vessels, multiple small angiomatous changes in the iris, luxation of the lens into the anterior chamber, and total detachment of the retina.

Although there is no clear evidence of heredity, there is general agreement that an hereditary tendency is often present. Werner (1952) thought that in the Sturge-Weber syndrome as in other phacomatoses an inherited weakness of the germinative plasma is responsible for a developmental inhibition of the as yet undifferentiated neuro-epithelial cells, with subsequent damage to the skin and nervous system; there is certainly also a disturbance of the mesodermal structures. The inhibitory factor is unknown, but Werner mentions rubella embryopathy, and suggests that the various phacomatoses depend on the manner and scope of the inhibition, on the time of its action, and on the degree of inherited tendency. King and Schwarz (1954) also favoured embryonic maldevelopment, and quoted Sabin (1917) and Streeter

(1918), who postulated a developmental vascular inhibition as the causative factor.

Bock (1950) reported an unusual case of the Sturge-Weber syndrome in a child with bilateral glaucoma, haemangioma of the face and forehead, cerebral changes, and epileptic fits. The ocular tension rose after massage but was little influenced by homatropine.

It is not clear how the glaucoma arises. The abnormal condition of the choroidal vessels and frequent presence of angiomata and intra-ocular capillary dilatations lead to venous stasis and disorganization of the circulation of fluid. Deficient drainage, such as is found in buphthalmos without naevus flammeus, and developmental anomalies (*e.g.* absence of the canal of Schlemm) may also play a part. Duke-Elder (1954) wrote that, although glaucoma-like naevus is congenital and buphthalmic in type, cases of simple, chronic glaucoma without deformation of the globe do occur. This glaucoma is often chronic and non-inflammatory, associated with deeply cupped and atrophic discs and progressing to complete blindness, though many patients may keep their sight for a long period despite increased ocular tension.

As the site of the brain changes is far removed from the motor region, the epileptic fits are probably due to the consecutive shrinkage of the grey matter. The angiomatous appearance of the cerebral vessels is frequently seen in younger patients, and the atrophy and calcifications in those of more advanced years, although pronounced calcifications have been found in a child aged 14 months, and no calcium shadows in a man aged 45. The oldest case recorded seems to be that of Frigyer and others (1953). A man aged 63 had naevus vasculosus on the left side of the face, glaucoma of the left eye, right-sided spastic hemiparesis, epileptic fits, and mental debility.

My patient displayed symptoms of incomplete Sturge-Weber syndrome, with widespread naevus flammeus, more pronounced on the left side of the face and trunk, and left-sided glaucoma with proptosis. As no *post-mortem* examination was made, nothing can be said about the condition of the cerebrum.

Cerebral Melanoma.—The combination of Sturge-Weber's disease with melanoma of the brain has not, to my knowledge, been reported before. The melanoma in the case described above was thought by one neuro-surgeon to have originated in the eye, but this suggestion could not be proved because no *post-mortem* examination was carried out. It may even have originated in an insignificant overlooked lesion of the skin, especially since the patient had been suffering from widespread disturbance of the cutaneous pigment. Any atypical or bizarre malignant skin neoplasm may be a malignant melanoma. Allen (1949) pointed to the presence of clusters of altered cells in malignant pigmentation, and Cade (1957) has stressed the importance of this point in diagnosis.

No such alterations were found, however, in the histological examination of my patient. In his case the melanoma could have developed independently

of either the eye or the skin, from pigmentary changes in the meninges themselves. Neither eye showed, until his death, the slightest evidence of being affected by primary melanoma. No cutaneous naevus pigmentosus was seen. Both local relapse and metastasis to the liver started some months after the operation on the brain tumour, which suggests that the secondary deposits originated in that tumour.

Touraine (1949) regarded the tendency to metastasis as one of the features common to both cutaneous melanomata and those of the central nervous system (Pol, 1905; Boit, 1907; Ehnmark and Jacobowsky, 1926; Baumecker, 1929; Foot and Zeek, 1931, Case 2). He considered that malignant melanomata usually developed from naevi pigmentosi or from precancerous melanosis, but might also originate in an apparently normal skin. This also applied to malignant melanomata of the meninges, which might develop from melanism or melanotic patches, but also in a tissue, normal in appearance, containing disseminated melanocytes. He regarded the melanogenesis and the subsequent pigmentation as a normal process of ectodermal origin, common to the skin, some mucous membranes, and the soft meninges.

Willis (1952) thought that interpretation of the origin of the growth was open to grave doubts, unless the cases reported by Hassin and Bassoe (1929) and Foot and Zeek (1931, Case 2), in which metastatic melanotic growths accompanied the allegedly primary melanotic tumours of the meninges or nervous system, were accepted. However, there is no intrinsic reason why malignant melanomata of the brain should not give rise to metastases, although so far no indisputable examples of this occurrence have been noted, and a critical study of such cases, including careful necropsy, is necessary.

As no adequate investigations were carried out in the case described above, and the *post-mortem* findings are lacking, each of the theories regarding the origin of the brain melanoma put forward has its merits and defects, and none can be either fully approved or completely discarded. The most acceptable is that of the simultaneous coexistence of various melanotic anomalies due to generalized pigment disturbance. As far as the melanotic changes are concerned, it appears to me that my patient's symptoms might be regarded as a supplementary extension to Touraine's original description of *melanose neurodermale*, presenting instead of cutaneous melanosis, amelanotic changes in the skin, melanoma of the brain (parts of which were entirely amelanotic), and also melanosis of the sclerae. On admission, the case was thought to be a link between Sturge-Weber's and von Hippel-Lindau's syndromes, on account of the angioma-like vessel in the right fundus. In the light of the evidence, it should rather be considered as a link between Sturge-Weber's syndrome and Touraine's *melanose neurodermale*.

Summary

A case is presented in which glaucoma of the left eye was combined with naevus flammeus of the face and trunk and generalized disturbance in

pigment distribution, culminating in the development of a brain melanoma.

Pear-shaped pupils, high-arched palate, and dental abnormalities were secondary features.

The case may be regarded as an incompleted example of Sturge-Weber's syndrome coexistent with the melanotic dysplasia described by Touraine as *melanose neurodermale*.

I wish to express my thanks to Colonel Tristram Samuel for his kind encouragement and permission to publish the case, to Professor Willis and Dr. Bickerstaff for their kind help and advice, and to Miss Campbell for the drawing of the pear-shaped pupil.

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EXAMINATION OF THE EYE WITH INFRA-RED RADIATION*

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Photography with Infra-Red Film

Dekking (1933, 1934) published several photographs of eyes with dense corneal opacities using infra-red film and radiation of a wave-length of about 10,000 Å.U. His photographs strikingly demonstrated how infra-red radiation could penetrate a dense corneal opacity, and clearly record the appearance of the iris and pupil, which were invisible by ordinary methods of observation and photography. Similar infra-red photographs were later published by Mann (1935) and Feldman (1936). Kugelberg (1934) attempted infra-red photography of the fundus, but failed because of the difficulty in obtaining contrast and detail.

Photography and Direct Observation with the Infra-Red Image Converter Tube

Zworykin and Morton (1936) described an indirect method of infra-red photography which involved the use of an infra-red image converter tube. This electronic device consists of an evacuated tube, at one end of which there is a cathode coated with caesium silver oxide upon which the image of an object illuminated with infra-red radiation is focused. When a high electronic potential difference is applied between the cathode and anode, electrons are emitted from the cathode in proportion to the illumination at any point. The electron discharge on striking the anode at the other end of the tube, which is coated with a fluorescent material, produces a visible picture. This can either be viewed directly or photographed with a conventional film and camera (Fig. 1, opposite).

The infra-red image converter tube has the following advantages over infra-red film in the examination of the eye:

(1) It dispenses with the necessity for photography and infra-red film.

(2) The image can be focused directly on to the anode (with infra-red film the image is invisible and focusing must be calculated).

(3) Moving phenomena, such as pupillary activity and movements of the eye itself, may be directly observed in darkness without the aid of infra-red cinematography.

(4) The converter tube is sensitive to infra-red radiation of longer wave-length than infra-red film: such longer infra-red rays more readily penetrate opaque media.

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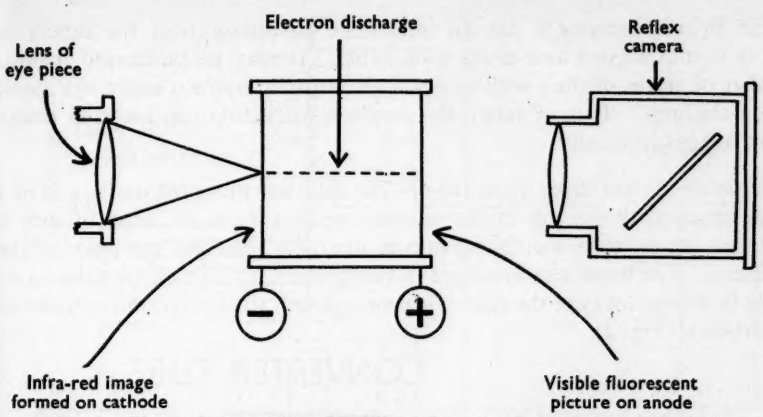

FIG. 1.—Electronic conversion of invisible infra-red radiation into a visible image.

Vaško and Peleška (1947) photographed the iris and pupil through an opaque cornea with a German army-type tube. Peleška (1951) described how fresh corneal infiltrates were more transparent than old opacities, how atrophied iris epithelium appeared black and could be mistaken for a coloboma, and how the opaque lens was more translucent than in normal light, but there was no possibility of examining a fundus through a cataract.

Kelly (1948) examined the anterior segment with the British army-type tube which was mounted on a slit lamp. He found the method of value in assessing the advisability of corneal grafting in certain cases. Marshall (1953), using a similar tube, observed eye movements away from fixation in rod scotometry. Knoll (1953) used the American tube for determining pupillary size and the refraction of the eye at low levels of illumination. Chin and Horn (1956) used an infra-red image converter tube to measure changes of refraction in dim illumination and in darkness.

COMPARISON OF INFRA-RED IMAGE CONVERTER TUBES NOW AVAILABLE

The value of an examination of the anterior segment of an eye with a completely opaque cornea before corneal grafting and the appearance of pigment lesions when viewed by infra-red radiation stimulated the author to compare the various types of infra-red image converter cells at present available. It was not possible to obtain a suitable instrument from the manufacturers of electronic tubes in Great Britain, and the investigation was limited to the American "I.P.25", German "Vampire", and British "R.G." army-type tubes. Both the American and the German tubes are focused electromagnetically. The former gave a small bright image, but it was difficult to photograph. The latter produced a larger image, but unfortunately gunsight wires in the tube interfered with photography. Both are very difficult to obtain in Great Britain, and the American tube is very expensive.

The British army-type has an immediate advantage over the others in that it is inexpensive and easily obtainable. It may be purchased from a number of stores dealing with ex-army electrical equipment and costs about fifteen shillings. It is of relatively simple construction, an inverted image being focused optically.

Use of the British Army-Type Tube.—The tube was mounted on the end of a beam attached to the top of the microscope of a Zeiss slit lamp in such a way that the cathode was facing and in line with the right eye-piece of the slit lamp. The beam was arranged to swing in an arc, so that the tube could easily be moved away to the right when an ordinary slit-lamp examination was undertaken (Fig. 2).

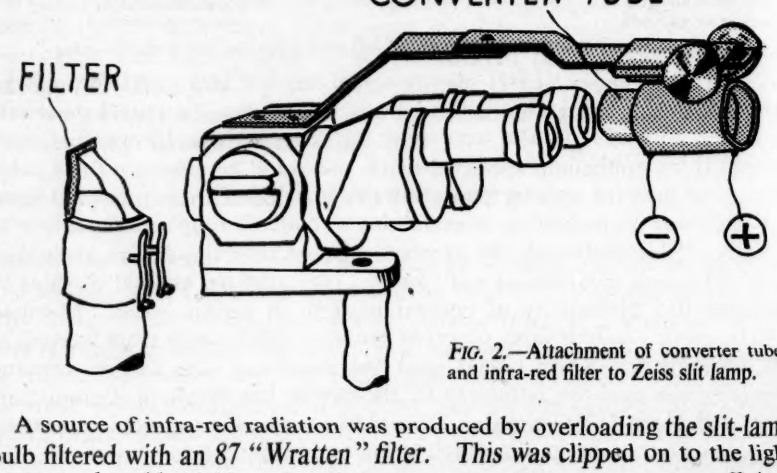

FIG. 2.—Attachment of converter tube and infra-red filter to Zeiss slit lamp.

A source of infra-red radiation was produced by overloading the slit-lamp bulb filtered with an 87 "Wratten" filter. This was clipped on to the light source and could quickly and easily be removed. The tube was most efficient when operating at 8,000 volts. Fig. 3 (opposite) shows the circuit of the power unit employed.

Results obtained with the British Army-Type Tube.—For the patient the only sensation was one of slight warmth on the eye. For the examiner the use of the instrument was no more difficult than an ordinary slit-lamp examination. The image formed on the anode was sufficiently bright for a detailed examination to be made with ease. Photographs of the image on the anode were made with a single lens reflex camera (a reflex Korelle), but exposures of about one half of a second were required, even with fast film.

Fig. 4 (opposite) shows a normal partially dark-adapted eye photographed through the tube. The grid wires on the anode can be seen.

The only corneal lesion which proved to be completely opaque to infra-red radiation was calcareous degeneration. With the overloaded slit-lamp bulb

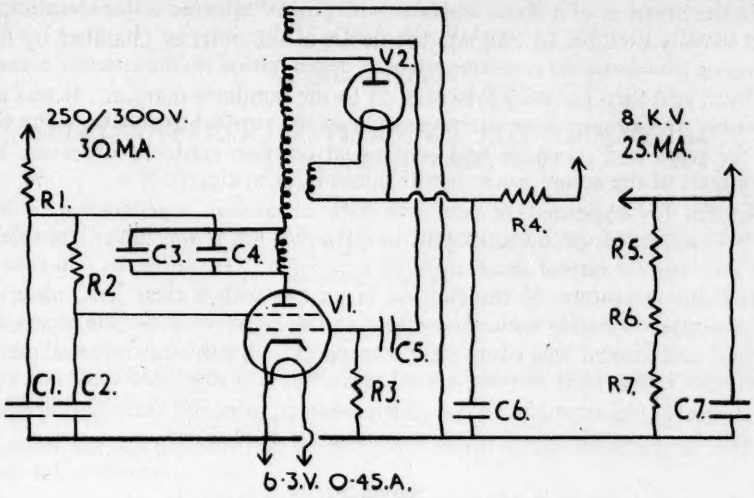

FIG. 3.—Circuit of power supply.

Value of Components

V.1. 6V.6.G.T.
V.2. U.37.
R.1. 470½ W.
R.2. 33 K.½ W.
R.3. 47 K.½ W.
R.4. 220 K.½ W.
R.5., R.6., R.7., 33 m. high voltage type.
C.1. 0·1 mfd.
C.2. 0·02 mfd.
C.3., C.4., Total of 0·001 to 0·0015 mfd. S.M.
C.5. 0·001 mfd. S.M.
C.6., C.7., 0·001 mfd. 10 U.V. Wkg.

all other opacities were penetrated, although, the greater the vascularity of the opacity, the greater the transparency. Corneal opacities which were penetrated by infra-red radiation in life were found to be opaque within a few hours of the enucleation of the eye.

FIG. 4.—Normal partially dark-adapted right eye photographed in darkness through converter tube (two sources of infra-red radiation were used).

In the presence of a dense leucoma with partial calcareous degeneration, it was usually possible to estimate the depth of the anterior chamber by first bringing into focus the area of calcareous degeneration on the anterior corneal surface, and then focusing forwards on to the pupillary margin. It was not possible to form any accurate estimation of the corneal thickness. The size of the pupil and its shape and consensual reaction could be observed, but very little of the actual iris structure could be seen clearly.

Except for hypermature cataracts with calcareous degeneration, a lens opacity appeared quite transparent to infra-red, but it was never possible to see any normal retinal detail through a cataract. However, in one case of malignant melanoma of the choroid in an eye with a clear lens, observed through the converter tube, the tumour stood out black while the secondary retinal detachment was completely transparent. In this case transillumination with visible light showed a solid mass, but it is suggested that, in a case of suspected melanoma in an eye with a cataract, infra-red examination might be helpful when the result of transillumination is in doubt.

Summary

The earlier work with infra-red film is reviewed. The advantages of the infra-red image converter tube over infra-red photography in the examination of the anterior segment in the presence of a corneal opacity are described. The various types of converter tube are compared and the advantages of the British army-type tube discussed. Such a tube, mounted on a slit lamp, is of value in assessing the state of the pupil and the depth of the anterior chamber before corneal grafting. The tube may also prove to be useful in diagnosing choroidal melanoma in an eye with a cataract, which prevents conventional observation of the fundus and in which transillumination with visible light gives equivocal results.

I should like to thank Mr. E. F. Fincham for his help and advice, Dr. P. Hansell and his staff for preparing the illustrations, Dr. H. J. A. Dartnall for lending an American converter tube, and Dr. R. Barer for lending a German model.

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Brit. J. Ophthal. (1958) **42**, 311.

CASE NOTES

SPONTANEOUS HYPHAEMA WITH ABNORMAL IRIS VESSELS*

BY

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A **woman aged 42** was examined in the Casualty Department of the Royal Eye Hospital, London, at 8.30 p.m. on June 30, 1956.

She complained of slight blurring of right vision associated with a little pain which had begun 7 hours before.

This had never occurred previously. There was no history of physical strain or excitement, nor was there any relevant family or personal history concerning her ocular or general condition.

Examination.—Visual acuity was 6/9 in the right eye and 6/6 in the left eye uncorrected.

External examination of the right eye showed no abnormality. A fine stream of blood was squirting without pulsation into the aqueous humour from a small ovoid sac attached to the pupillary margin at 11 o'clock. This well-defined stream descended in the aqueous humour to form a thin layer of blood in the lower part of the anterior chamber. Although the stream was continuous the amount of blood in the anterior chamber did not increase, suggesting that it was being removed from the angle as quickly as it was collecting. Occasional slight irregularities in this flow caused "mushrooming" in its course (Figure).

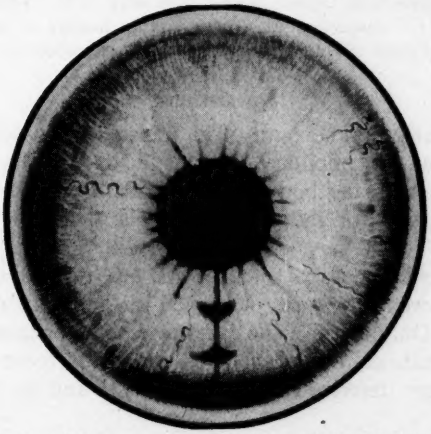

FIGURE.—Anterior segment of right eye. A small stream of blood is being ejected from a micro-aneurysm into the aqueous humour, "mushrooming" in its course and forming a thin hyphaema. Tortuosity of iris vessels is seen especially at 9 o'clock. The iris stroma is rather more loose than is apparent in the illustration.

* Received for publication April 12, 1957.

Slit-lamp examination showed that the blood was being ejected from a small pale pink sac on the pupillary margin, the appearance of which strongly suggested a ruptured microaneurysm. Its size was about 0·15 mm. There was a similar but smaller intact swelling at 1 o'clock on the margin.

The iris stroma was grey and loose and the iris vessels were clearly visible except at the sphincter. They were tortuous and slightly distended, especially one vessel in the 9 o'clock radius. Their appearance, in fact, resembled an acute inflammation, but there were no other signs of an inflammatory condition. At 10 o'clock on the surface near the margin there was a fine network of capillaries which showed the "cattle-truck" phenomenon.

The pupils were both approximately 2 mm. in diameter and their reactions were normal.

The right lens and vitreous were normal. The fundus showed a small scar of old choroiditis near the disc which corresponded with a small steep-edged scotoma.

The left eye was normal apart from the iris, which showed the same scarcity of pigment and loose fibrillary structure as the right. The vessels in the iris stroma showed more marked tortuosity than in the right eye, especially those in the 6, 9, and 11 o'clock radii, and gonioscopy revealed the 6 o'clock-vessel to emerge from a small conglomeration of loops in the angle of the anterior chamber. There was a small swelling on the pupillary margin at 1 o'clock similar to those in the right eye.

A placebo was prescribed and the patient was asked to return on the following morning when a small thrombus was seen attached to the sac at 11 o'clock in the right eye. One day later this thrombus had disappeared and all other signs of blood in the anterior chamber were gone. The vision in this eye was still 6/9 (corrected to 6/5) but no longer subjectively blurred.

A full investigation of the respiratory, digestive, and central and peripheral nervous systems showed no abnormal findings; blood examinations (including clotting, prothrombin, and bleeding time, and capillary fragility) and urinary examinations were normal. In liver function tests, serum bilirubin, direct and indirect, alkaline phosphatase, and zinc sulphate turbidity were all within normal limits. At the first estimation, thymol turbidity showed 2 units and thymol flocculation was + +. The second estimation showed 4 units and + + respectively. (There was no history of hepatitis or jaundice and the significance of these slightly abnormal findings was never discovered.)

Discussion

This case is reported on account of its unusual iris vascular pattern with micro-aneurysms at the pupillary margins. No vascular anomalies were discovered elsewhere in the body and no disease processes were found to suggest an explanation for the unusual vessels. One therefore assumes a congenital origin.

Angiomatous malformations of the iris are rare (Duke-Elder, 1940). Rodin (1929) reviewed nine cases of alleged iris angioma and described one case of his own. Only three of the cases reviewed appeared to him to have been truly angiomatous, but all showed tumorous formation. This also applies to the cases described by Fuchs (1913) and to that reported by Knapp (1928).

The iris vascular pattern of the case presented here showed no indication of neoplastic origin. The tortuosity of the iris vessels, the loose fibrillary structure of the stroma of the iris, and the scarcity of iris pigment resembled the findings in the case reported by Tyson (1932), but the latter also

presented a naevus flammeus of the face and globe associated with glaucoma and a calcified vascular growth of one occipital lobe of the brain with homonymous hemianopia on the contralateral side. As mentioned above no such signs of systemic abnormalities were found in the new case described in this paper.

Summary

A patient is described in whom the iris vascular pattern showed dilatation and tortuosity of some of the larger vessels. These abnormalities were associated with a fine capillary network near the right pupillary margin, and at least one micro-aneurysm on the right pupillary margin which ruptured and ejected a fine stream of blood into the aqueous humour.

I am indebted to Mr. T. M. Tyrrell for permission to publish this case, to Mr. T. A. S. Boyd for his help with preparing the text, and to Mr. T. Tarrant for his skilful preparation of the illustration.

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Brit. J. Ophthal. (1958) **42**, 314.

INSECT STING OF THE LID—AND A SEQUEL*

BY

FRANK W. LAW

London

AN anxious parent brought her 6-year-old son to me 2 or 3 years ago because he had been "stung on the eye by a bee". A cheerful and co-operative child showed me an irritable watery eye with some injection. It was quite definite that the insect had attacked the upper lid at about its mid point, for the site of puncture had been quite obvious at the time; no sign of this remained by the time the patient reached me 2 days later. There was no corneal stain, nor any sign of abnormality to be discovered on the upper tarsal conjunctiva—certainly no sting nor puncture hole. I was at a loss to explain the occurrence of injection and irritation from what I then thought to have been no more than a skin lesion; I prescribed some simple drops and asked that the patient should be brought back in 2 days' time.

On his return the eye was as irritable as ever, and fluorescein staining and examination under the slit lamp revealed a multitude of the finest imaginable linear or slightly curved scratches. I had never seen such a delicate tracing. Eversion of the upper lid revealed, after close inspection, a slightly raised central area, a little redder than the rest of the conjunctiva; by manipulation of the lid after cocainization of the eye the business end of a sting was made to extrude itself; this was grasped with forceps and withdrawn. It is clear that the sting pierced the tarsal plate from the skin surface and intermittently protruded so as to touch the cornea—for this was non-staining at the time of the first visit, though the eye had obviously suffered recent irritation.

The sting on examination proved to be indeed that of a honey bee; the characteristic barbs are unmistakable, and are shown here in surface and side view (Figs 1 and 2). These barbs retain the sting in position and

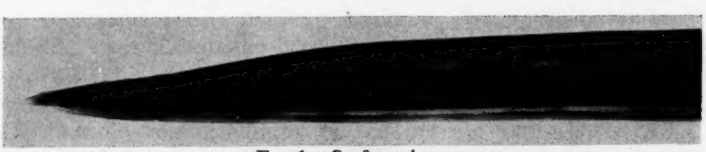

FIG. 1.—Surface view.

FIG. 2.—Side view.

* Received for publication July 25, 1957.

indeed assist its deeper penetration even when the bee has departed; reflex movements of the lancets continue and poison continues to be injected into the wound for some time afterwards if the sting is allowed to remain. This is a most interesting phenomenon, easily confirmed, which I have many times experienced.

The eye recovered from the moment of removal of the sting, and I have not heard of the patient again.

* * * * *

Bell and Weir (1947) read a paper before the Ophthalmological Society of the United Kingdom at the Congress held in Glasgow under the presidency of the late Professor Ballantyne, in which they set out to answer the question:—why do we not see blurred images on transferring our fixation from one point to another? Their thesis was illustrated by an ingenious experiment erected before the audience; members had to transfer their gaze from a right-hand fixation point to a left-hand fixation point, between which there was a patch of light flickering at 120 times per second. A series of bright patches was seen extending right and left to a different extent with different observers.

The paper was interesting and so was the discussion. My contribution to this was to say that, when we make horizontal glancing movements of the eye, other than the very shortest, we habitually close our eyes during the movement, so the question raised was quite academic; blurred images were not seen because the eyes were closed. To me this was a matter of common observation; the eyes are as it were "swept across" by a blink. Further subsequent observation has done nothing to alter my contention; and I was astonished to find at the meeting that—as the account says—"some dubiety of opinion was expressed by the audience on this point". It is perfectly possible to swing one's gaze across with the eyes open, as a conscious effort; but the *natural* movement is accompanied by a blink.

* * * * *

Immediately my small patient had left my house for the second time, relieved of this sting, my thoughts turned to this debate in Glasgow of some years before—for *all the fine scratches on his cornea were horizontal* or nearly so, some with a slight curvature. This appeared to suggest that the orbicular action in glancing movements is firmer than that employed during the simple blink reflex with the eye stationary. I do not know whether that is a fact, or, if it is, whether physiologists are aware of it, and of its significance.

What interested me even more, however, was that it appeared to prove in a dramatic and conclusive manner my contention that the lids close during glancing movements.

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Brit. J. Ophthal. (1958) **42**, 316.

CORRESPONDENCE

PRESERVATION OF CORNEAL GRAFTS BY FREEZING

To the Editorial Committee of the BRITISH JOURNAL OF OPHTHALMOLOGY

SIRS,—Mr. Rycroft, in his interesting article "Three Unusual Corneal Grafts" (Rycroft, 1957), describes a successful case of perforating keratoplasty using donor material which had been stored for one month in the deep-freeze at −79° C. He states that this establishes *primâ facie* evidence that long-term storage at low temperatures is possible, that modification of the deep-freeze technique has given encouraging results, and that in his opinion the deep-freeze method will become the bank method of the future.

Whilst agreeing on the future possibilities of the deep-freeze method for the preservation of corneal tissue, I believe that the present state of our knowledge regarding the use of such material should be clearly defined. Eastcott, Cross, Leigh, and North (1954), following the work of Polge, Smith, and Parkes (1949), investigated the preservation of corneal tissue by freezing and recorded a series of five lamellar and seven full-thickness grafts using tissue which had been stored at −79° C. after immersion in 15 per cent. glycerol saline. All the lamellar grafts were entirely successful. Each of the full-thickness grafts was associated with considerable post-operative opacification which cleared slowly, progressing to complete clarity in only one case. As a result of these findings the authors concluded that, whereas donor material from the deep-freeze was suitable for lamellar grafts, its use could not be advised in perforating grafts. Subsequent observers have confirmed these results using similar and modified techniques of freezing (McNair and King, 1955; King, 1957).

Mr. Rycroft may well have solved this difficult and intriguing problem, and we look forward with great interest to the publication of his results. It is pertinent to observe, however, that in the case described Mr. Rycroft appears to have relied upon the protective action of 15 per cent. glycerol saline, a method which does not differ in principle from that used by Eastcott and others (1954); therefore, until such time as we are presented with new evidence, it should be stressed that the use of such material for routine perforating keratoplasty is entirely unjustified.

Yours faithfully,
A. G. LEIGH.

100 HARLEY STREET,
LONDON, W.1.
29 January, 1958.

To the Editorial Committee of the BRITISH JOURNAL OF OPHTHALMOLOGY

SIRS,—Whilst the routine use of deep-freeze corneal graft material for all penetration grafts is not yet feasible, only the constant investigation and trial in selected cases under suitable conditions, with the accurate assessment of results, has justified future optimism for this method of preservation. Eastcott, Cross, Leigh, and North (1954) reported seven cases of human penetration grafts in which the grafts had been preserved by the deep-freeze method; two of these were stated to be completely successful, three were partially successful with no deleterious influence on vision, and there were two failures which were subsequently regrafted with success. These authors stated that the method had been found by them to give satisfactory results in the preservation of human corneal tissue for grafting.

In ophthalmology the future promise of the glycerol-saline method of preservation at −79° C., first described by Polge, Smith, and Parkes (1949), rests on two established facts:

(1) Lamellar grafts have proved to be entirely suitable when preserved in this fashion (Eastcott and others, 1954; King, 1957).
(2) At least four successful cases to date of penetration grafts preserved in a similar manner have been carefully reported and followed up for many months after operation (Eastcott and others, 1954; Iliff, Wood, and Hollander, 1956; Leigh, 1954; Rycroft, 1957).

Nor does it follow that the opacification of the failures is necessarily due to the specific method of preservation, since many factors are involved and opacification can occur from these various causes in every method of preservation. For example, in the series previously quoted, one deep-freeze graft which had failed had been carried out in an eye in which a previous fresh graft had also failed "in the absence of any recognized post-operative complication". But the failure of the deep-freeze graft in this case was due to a condition which can occur in any type of penetration graft, namely the development of a posterior graft membrane; the graft tissue itself remained clear. Nevertheless, it is known that the behaviour of a deep-freeze full-thickness graft is distinctly different from that of a fresh graft and is associated with more prolonged oedema; this is a problem which remains to be solved.

The clinical approach to the solution must be further study of many selected cases, with variables controlled as far as possible in relation to the initial lesion, the type, age, and density of the scar, the degree of vascularization, the relationship of host and donor thickness, the age, sex, and general condition of the patient, the standardization of surgical and nursing technique, and the strict control of the sterility and method of donor collection. It is equally important that caution should also be exercised in the interpretation of the cases of failure in assessing the exact role which the method of preservation plays.

The essence of the laboratory problem is the behaviour of the endothelium and Descemet's membrane under the thermodynamic and biological stresses of low temperatures with subsequent reconstitution. Mr. Leigh suggests that the problem may have been solved, but I am sure he will agree that a solution is unlikely to be found in the operating theatre, but more likely in the laboratory; the ophthalmologist is only the builder and the biologist is the real architect.

Studying this particular problem, Ridge (1956) has shown that at pH 8·8 the rabbit endothelium is completely resistant to the toxic action of 10 per cent. glycerol, and that under these conditions the endothelium is completely protected against the harmful effects of freezing and thawing. At Bethesda, Draheim, McPherson, Evans, Perry, and Earle (1955) stated that preliminary tissue culture viability tests indicated that the rabbit cornea retains viability when preserved by this method.

At East Grinstead we are fortunate to work under the guidance of such authorities as Drs A. S. Parkes, Audrey U. Smith, and R. H. Billingham, and their valuable advice controls our deep-freeze investigations. They have pointed out the importance of such factors as the variability of temperature in a deep-freeze bank, the risk of carbon dioxide diffusion into any glass container which is not effectively sealed, the need for a slow rate of freezing and rapid thawing, and also that even yet the optimum percentage of glycerol is in doubt as well as the adequate time of soakage.

Surely, therefore, it is reasonable to expect that this problem will eventually be solved by the united efforts of the biologist, ophthalmologist, and pathologist, and that it will be possible to establish in every country of the world, where keratoplasty can alleviate blindness, Regional Banks in which adequate supplies of sterile and suitable corneal

donor material are readily available at the precise time when they are required. This is the objective of the investigations of the deep-freeze method of preservation.

Yours faithfully,

B. W. RYCROFT.

35 HARLEY STREET,
LONDON, W.1.
18 February, 1958.

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BOOK REVIEWS

Reactive Cellular Changes in the Cornea and Retina. (Reaktive Zellveränderungen in Hornhaut und Netzhaut.) By H. Pau. 1957. Samml. zwang. Abhild. Geb. Augenheilk., No. 13. Pp. 111, 100 figs, 82 refs.

This publication combines two separate studies under one general title. The first section deals with the reactions to injury and the regeneration of the cells and tissues of the cornea, and the second describes histological findings and other observations in retinal tears and detachments and places less emphasis on cellular reactions.

Most of the information on corneal cellular reactions has been derived from the rabbit cornea after experimental lesions and, whilst it is clearly the main intention of the author to describe the histological findings, it is regrettable that only the briefest mention is made of experimental procedures. However, the corneal changes after injury are described carefully in an interesting manner and with the aid of microphotographs which almost always illustrate well the points mentioned in the text. In many instances, a silver-staining technique was particularly useful for displaying details of nuclear structure. Frequent reference is made to corneal parenchymal cells which (as the author explains in a footnote on page 21) are not analogous to the parenchymal cells of other organs but are specialized connective tissue cells of the substantia propria. Small chromatin granules, usually five in number, were demonstrated in the nuclei of the so-called "fixed" cells of the cornea, and the important role of these cells in the healing of corneal wounds is described. It is claimed that the leucocytic cells in some forms of keratitis originate from corneal cells, the chromatic granules enlarging and becoming connected by bridges of nuclear material to form the segmented nuclei typical of some leucocytes. Similar changes were found in regenerating epithelial cells which proliferate and migrate to cover superficial lesions in the early stages of the healing process. The parenchymal cells of the cornea are also involved in the vascularization of corneal lesions and in the regeneration of corneal nerves, undergoing transformation to vascular endothelial cells and replacing Schwann cells.

The section dealing with retinal detachments contains descriptions of the histological

appearances in human eyes of adhesions between the retina and condensations in the vitreous body, and of the associated sclerosed area of the retina. The cellular reactions in the neighbourhood of the vitreo-retinal adhesions consist of hyperplasia and migration of the cells of the pigment epithelium and proliferation of vascular connective tissue. All the microphotographs illustrating the findings in retinal tears and detachments have been placed at the conclusion of the second section; this is disappointing because the reading of the section on the cornea is greatly facilitated by a good arrangement of the illustrations, most of which appear close to their references in the text.

Traitement chirurgical des affections oculaires, Vol. 1. By L. Guillaumat, L. Paufique, R. de Saint-Martin, S. Schiff-Wertheimer, and G.-P. Sourdille. 1957. Pp. 440, 105 illus. Doin, Paris. (6,500 *Fr. frs.; 6 guineas*).

The technique of surgical operations in the field of ophthalmology has been covered by a number of text-books written in recent years, and in most of them operative procedures are given in detail, and the numerous types of operation for each particular indication for surgery are described.

The first volume of this new work is unusual in many respects. It is the work of a number of authors, each of whom has written his own sections of the book, and all of whom have reviewed the entire book. This has not deprived the author of any particular chapter of his opportunity of expressing his own personal preferences, but has enabled him to do this and at the same time to present any such preferences in their proper perspective.

Recognizing what is too often forgotten, that it is when to operate and which operation to perform for the case under consideration, that requires the thought and guidance of the surgeon, the authors have given full consideration in each section to the indications for operation, and to the features which determine the most appropriate moment for surgical intervention.

Operative technique is given its proper place, and the advantages and disadvantages of many modifications of standard surgical procedures are discussed in such a way as to enable the reader to think for himself, and to form his own opinion. The authors carry their understanding of the needs of the reader yet further by each discussing in the latter part of his own section the normal convalescence to be expected following an operation, and the various complications to which it is liable and their treatment.

The first volume deals with cataract, glaucoma, iridocyclitis, and traumatic surgery, and includes an introduction covering the layout of the operating theatre, its illumination and equipment, and general notes on pre-operative preparation and medication, and on local and general anaesthesia. The second volume will include sections on corneal surgery, retinal detachment, enucleation and exenteration, tumours of the globe and ocular adnexa, plastic surgery of the lids and orbit, squint, and ocular palsies.

The success with which the authors have combined a book on operative technique with so much valuable information about the pre-operative management, the operative mishaps which may occur during the operation, and the treatment of the complications, has resulted in a volume which should be of equal value to the student of ophthalmic surgery and the established surgeon. The former is taught to think in such a way that he can decide for himself the course to follow, and support his decision by logical argument, and the latter may learn a great deal about how to avoid some of the commoner errors of technique, and how best to treat the complications which unhappily befall even the most competent of operators. It is with pleasant anticipation that one looks forward to the publication of the second volume. The regret expressed in the foreword by the remaining authors at the loss of their colleague, Gabriel-Pierre Sourdille, will be felt fully by all those who have known him, and with whom he has shared his wide experience of the problems of ophthalmic surgery.

Slit-Lamp Gonioscopy. By G. Gorin and A. Posner. 1957. Pp. xii+176, 1 col. pl., 67 figs, 142 refs. Baillière, Tindall and Cox, London. (56*s*.)

This book is a short, simple, and yet comprehensive manual of gonioscopy. The opening chapters deal with the anatomy of the anterior chamber and of the drainage channels, the history of gonioscopic investigation, and technique; this last is based on the use of the Goldmann type of lens with a Haag-Streit or Zeiss type slit lamp, and as described is eminently suitable for routine use in consulting room or out-patient work. The gonioscopic appearances are then described in normal cases, in glaucoma of various types before and after operation, in congenital anomalies, and in those conditions—injuries, foreign bodies, neoplasms, and the like—which may implicate the region of the angle. Other clinical aspects and the help which gonioscopy may give in the management of the case are suitably indicated, and the textual description of the various conditions is admirably supplemented by many half-tone illustrations. The book is excellently produced and can be thoroughly recommended to those wishing to learn the technique, interpretation, and value of gonioscopy as a routine procedure.

A System of Ophthalmic Illustration. By P. Hansell. 1957. American Lecture Series Publication No. 289, Pp. 114, 78 figs. (4 col.). Thomas, Springfield, Ill.

The value of good photographic records in all branches of ophthalmology is undoubted, but even the most skilled photographer will not obtain satisfactory results unless the special problems of work in this field are appreciated. These problems are clearly explained in this book, which describes the essential equipment and materials, and working techniques, in studio, ward, operating theatre, and consulting room.

Motion picture photography and such special techniques as keratography, retinography, goniophotography, and slit-lamp photography are also discussed. The photography of pathological specimens and special preparations is briefly described and the value of co-operation between the ophthalmic artist and photographer is stressed.

Although this book is primarily intended for the professional medical photographer using specialized equipment, simple methods within the scope of the clinical ophthalmologist are also described by means of which routine records of average quality can be obtained. For this reason it is well worth reading by ophthalmologists to whom it will also give a clear idea of the possibilities and value of photographic records. The medical illustration departments of general hospitals will find this book of great help in producing a high standard of ophthalmic illustration.

The book is well written, clearly printed, and abundantly illustrated.

NOTES

Royal Eye Hospital

During the first week in February, 1958, a Refresher Course, held in the Courage Laboratory of the Royal Eye Hospital, was attended by fourteen ophthalmologists. The following subjects were dealt with:

Antibiotics, Genetics, Hypertension, Toxoplasmosis, Optic Atrophy, Contact Lenses, Visual Aids, Cortisone, Plastic Surgery, Sarcoidosis, Corneal Grafting, Lacrimal Sac Surgery, Glaucoma, Anaesthesia, Nature of Refractive Errors, Vertical Muscle Imbalance, Congenital Ocular Palsy, and Radiotherapy.

The course was so successful that it has been agreed to arrange a similar one in 1959. Ophthalmologists who are interested should write to:

G. W. Hoare, Holmeswood, Clytha Park Road, Newport, Mon.